Obstetric brachial plexus lesions

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Obstetric brachial plexus lesions

Aspects of diagnosis and treatment

ACADEMISCH PROEFSCHRIFT

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Willem Jan Roelof van Ouwerkerk

geboren te Rotterdam

promotor: prof.dr. W.P. Vandertop

Opgedragen aan
mijn ouders

Cornelia Elisabeth van Ouwerkerk – van Ooijen
en wijlen
mijn vader Roelof Lucas van Ouwerkerk

en

Dr. A.C.J. Slooff,
in leven neurochirurg,
pionier van de moderne behandeling
van het
obstetrisch plexus brachialis letsel
in Nederland en Europa

Contents

CHAPTER 1

INTRODUCTION

INTRODUCTION

An obstetric brachial plexus lesion (OBPL) is a stretch injury to the brachial plexus which occurs during delivery. Worldwide the incidence of OBPL in general shows a tendency to increase rather than to decrease over the past decennia. In the Netherlands the incidence has also increased but varies considerably depending on ethnical variance. Non-caucasian women have a higher risk for OBPL. In an area inhabited with many immigrants from Africa or Surinam, as in Amsterdam, the incidence over the past decennium was 0.46%, whereas the incidence in Caucasian women was 0.22% (thirty years earlier: 0.13%). Most publications seem to agree on obstetrical risk factors as maternal age over 35 years, multiparity, ethnic descent, diabetes, second stage of delivery of over one hour, breech presentation, operative vaginal delivery in multiparae and birthweight. Birthweight of over 4000 gram seems to be the most important risk factor for OBPL and moreover, birthweight is increasing over the years. However, as antepartum estimation of fetal weight is unreliable the occurrence of OPBL should be considered unpredictable. Recent studies concluded that, whatever risk factors present, prediction and consequent prevention of OBPL is not possible. Therefore OBPL will continue to demand medical attention in times to come.

Most infants recover spontaneously within weeks or months. Babies with severe lesions are at risk to develop serious functional limitations of the affected upper extremity and need specialized treatment by a multidisciplinary team. The damage to the peripheral nerve structures of the brachial plexus will cause varying degrees of muscle weakness in the affected shoulder and arm. These primary neurological lesions may lead to secondary deformities, that may even persist after neurological improvement, causing additional functional impairment. Neurosurgical treatment, with plexus exploration and reconstruction, is indicated when there is insufficient spontaneous recovery around the fourth month of life. Neurosurgeons are confronted with large neuromas after neurotmesis of plexial nerves as well as nerve root avulsions. Decisions as how to reconstruct a damaged brachial plexus and the chances of success of reconstructive procedures depend greatly on the correct diagnosis and intra-operative confirmation of the type of nerve lesion. Certain uncertainties still prevail in individual cases.

This thesis focuses on the state of the art regarding preoperative investigations and operative treatment in the current management of infants with OBPL.
The objectives of the thesis are

- to make an inventory of, and select, develop or modify ancillary investigative tools that will contribute to the most likely correct diagnosis of OBPL and possible secondary deformities,
- to evaluate the effect of a relatively easy to perform nerve transfer in carefully selected patients with a typical clinical picture lacking active shoulder exorotation,
- to improve the surgical technique to harvest the sural nerve, the most commonly used donor nerve, by developing a minimally invasive procedure.

After a general inventory of current practice and available preoperative investigations (chapters 2 and 3) the first objective was to reliably demonstrate cervical root avulsions with a new and easily applicable Magnetic Resonance Imaging (MRI) technique (chapter 4). Exact imaging of the course of nerve roots from the root entry zones in the spinal cord and further along their course within the spinal canal would have important surgical implications, as operative procedures to reconstruct the brachial plexus are based on nerve grafts from proximal cervical nerve roots to distal plexus structures. It is vital to know if a seemingly intact nerve root at its exit from a vertebral neuroforamen is proximally continuous with the spinal cord. Only then it has regenerative capacity and is suitable for grafting.
The second objective was to identify early secondary deformities of the shoulder in infants with an OBPL, who did not recover satisfactory within the first three months of life (chapter 5).
The third objective was to explain and effectively treat the persisting lack of spontaneous recovery of active shoulder exorotation in selected children with an OBPL, who showed satisfactory recovery of all other shoulder and arm functions (chapter 6).
The fourth objective was to improve the surgical technique to harvest a sural nerve, the most commonly used donor nerve in reconstructive peripheral nerve surgery, to avoid mutilating, large and potentially constricting scars in the skin of the calf (chapter 7).
Finally various aspects on diagnosis and treatment of OBPL are discussed and overall conclusions are formulated (chapter 8).

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CHAPTER 2

OBSTETRIC BRACHIAL PLEXUS LESIONS: AN OVERVIEW

Based on:

Management of obstetric brachial plexus lesions: state of the art and future developments.

Child's Nervous System 2000;16:638-644

Management of obstetric brachial plexus lesions: state of the art and future developments

ABSTRACT

Despite improving perinatal care the incidence of obstetric brachial plexus lesions (OBPL) has not declined. Most babies recover spontaneously. In 10–20% recovery is incomplete. To prevent lasting functional deficits early referral to specialized centers is necessary. If the biceps shows no function at 3 months, standardized clinical assessment and additional investigations must delineate the extent of a lesion. Detection of root avulsions by myelography and computed tomography combined with electrodiagnostics remains inconclusive in 15% of cases. Plexus reconstruction is performed during the 4^{th} – 6^{th} months. Contractures or deformities are treated conservatively or by orthopaedic surgery. Long-term rehabilitation is required. In future, aspects of prevention need attention. Improving imaging and neurophysiological techniques are promising for greater precision in detecting root avulsions and even spontaneous recovering nerves. Functional imaging will allow better understanding of central integration and plasticity. New pharmacological agents may promote nerve regeneration.

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State of the Art and Perspectives for the Third Millennium, Martinique,
27 November to 4 December 1999

Introduction

Probably due to an increase in birthweights the incidence of obstetric brachial plexus lesions (OBPL) of 0.5–3 in 1000 live births has not declined despite improved perinatal care [2]. Most infants (75–90%) recover spontaneously within weeks or a few months. In 20% there is no or incomplete recovery. Treatment of these patients is complex and requires a multidisciplinary approach. We discuss current management and future prospects.

Risk factors and etiology

Large, heavy babies (>4 kg) and small infants in breech presentation are predisposed to OBPL [2, 15, 18, 34, 38]. Other risk factors include shoulder dystocia, instrumental delivery, abnormal presentations, prematurity, and asphyxia. Maternal risk factors include diabetes, adipositas (pre)ecclampsia, and pelvic abnormalities [9, 38]. The risk of OBPL is increased in multiparous women and in women with previous such deliveries [1, 9]. The most common mechanism is a stretch injury to the plexus in cephalic presentation when shoulder dystocia requires extreme lateral flexion and traction on the head [35]. In breech delivery the spinal cord may additionally be displaced by longitudinal traction on the remaining head. The risk of even a bilateral lesion is increased in breech delivery [34]. Four basic nerve injury types have been described [33]. *Neuropraxia* is a temporary conduction block. In *axonotmesis* the axon is severed but surrounding neural elements are intact. In both types spontaneous recovery is likely. *Neurotmesis* is complete postganglionic disruption of a nerve. *Avulsion* is preganglionic disconnection from the spinal cord. The two latter types are managed by surgery.

Clinical assessment

The assessment and grading of functional status is difficult in babies. The use of different classification systems in the literature is confusing, such as those of Mallet [27], its modified version [14], the Medical Research Council Muscle Grading System or its modified version [17], the Narakas Classification of OBPL [29], and the Hospital for Sick Children Muscle Grading System [7]. For assessment the following classification is recommended [2, 29, 35]:

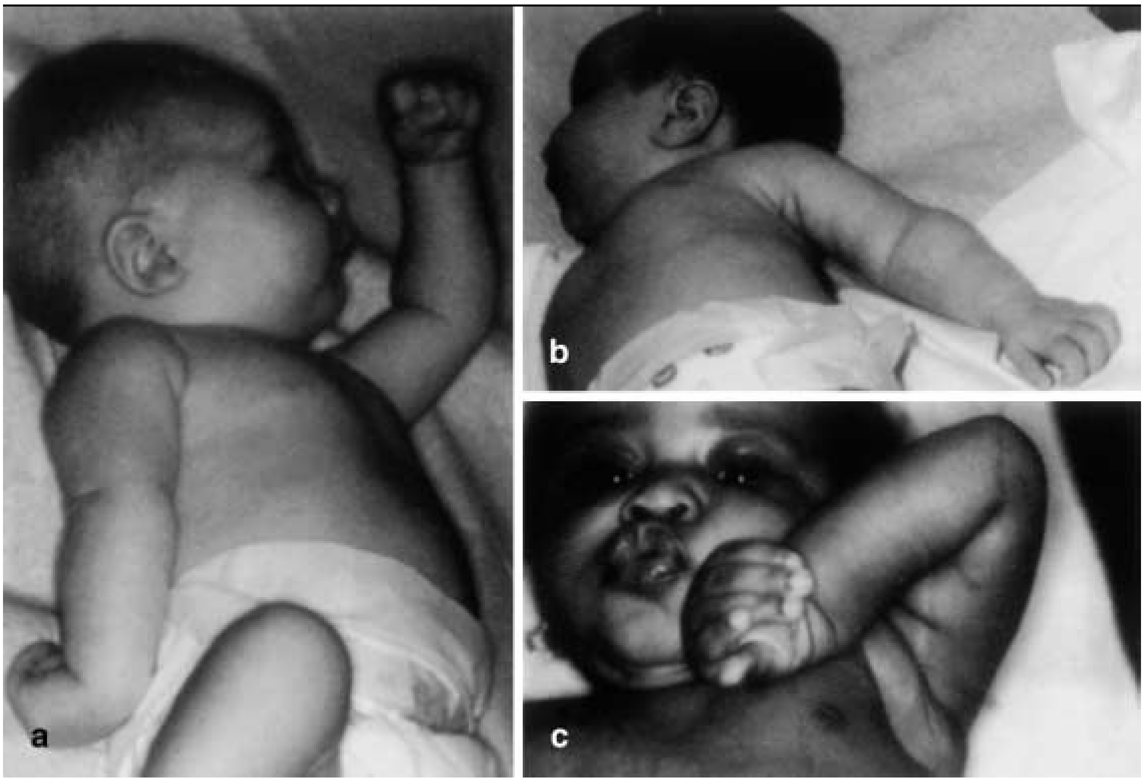
– Group 1: Lesion of the 5th and 6th cervical nerves or superior trunk with paralysis of shoulder abduction and exorotation, elbow flexion and forearm supination. About 90% of babies recover spontaneously.
– Group 2: As group 1 with an additional lesion of C7 or the medial trunk. The typical “waiter’s tip position” is due to paralysis of extension of elbow, wrist, and fingers (Fig. 1a). About 65% of cases show full recovery.
– Group 3: Except for some finger flexion paralysis is virtually complete. Horner’s sign is absent. Less than 50% of patients recover spontaneously.
– Group 4: Total plexus lesion with atonic, flail limb and Horner’s sign (Fig. 1b). Deficit is permanent. The “dominant C7 lesion” with paralysis of shoulder adduction and extension of elbow and forearm is subclassified here (Fig. 1c).

Consensus has been reached on the use of a practical grading system on outcome for shoulder, elbow, and hand function, as proposed by Gilbert and Raimondi [2, 7].
In OBPL the associated lesions include hematomas in the sternocleidoimastod muscle, fractures of clavicle, humerus, or even ribs, lesions of the phrenic, facial or hypoglossal nerves and even of the spinal cord [2, 35]. We have seen three tracheal ruptures. Babies with OBPL tend to turn their head away from the affected side (Fig. 1a, b). This may lead to shortening of the sternocleidoimastod muscle, torticollis, and positional occipital plagiocephaly.

Diagnosis and management

Diagnosis is usually straightforward. Rare cases of arthrogryposis, sepsis of the shoulder joint, fractures of clavicle or humerus, cerebral palsy, and cord injury should be differentiated from OBPL.
Physiotherapy should start not later than the third week. The chief aim is to prevent contractures and joint deformities. Parents are instructed to perform gentle but frequent exercises to maintain full passive exorotation and abduction in the shoulder, and extension in elbow, wrist and fingers. If spontaneous recovery does not take place within 2 months,

Fig. 1 a “Waiter’s tip” position in group 2 lesion. **b** “Total flail” arm in group 4 lesion. **c** Dominant C7 lesion



referral to a specialized center is recommended. *Follow-up* every 4 weeks in the first 3 months allows selection of babies with early recovery and good prognosis (about 80%) and those that should be surgically treated according to currently accepted criteria [2, 16, 35, 40]:

– Failure to recover biceps function, regardless of whether in combination with failure to recover extension of the wrist, fingers, and thumb within 3 months from birth
– Evidence of a severe lesion with Horner's sign, persistent hypotonic paralysis, persistent phrenic nerve paralysis, severe sensory disturbances, evidence of root avulsions on magnetic resonance imaging (MRI) or myelo-computed tomography (CT), persistent denervation in neuro-physiological studies

In babies meeting either of these criteria the microsurgical exploration of the plexus should not be postponed after the fifth month because the outcome worsens, with atrophy of sensory end organs and muscles, development of bone and joint deformities, and contractures by muscle imbalance.

Disintegration of central movement programs with time and neglect may adversely affect outcome [3, 22, 32].

Additional investigations in insufficiently recovering infants are centered on the issue of how to reliably detect complete or partial cervical nerve root avulsions and, in the latter case, how to determine the quality of a root for spontaneous recovery or its suitability for nerve grafting.

Imaging

Myelography and myelo-CT under general anesthesia may show pseudomeningoceles, subarachnoid space deformity, or missing root shadows (Fig. 2a, b) that have a strong association with root avulsions but do occur without or may lack surgically confirmed avulsions [5, 20, 30, 39]. Predicting a level of injury causes problems with contrast enhancing pseudomeningoceles obscuring multiple levels and also because of discrepancy between the location of root exit zones from spinal cord segments and that of respective intervertebral root exit foramina. Although MRI is non-invasive and performed under mild sedation imaging results with fast spin echo, three-dimensional MR myelography or MR neurography are promising but still lack resolution in small structures in infants or are technically too demanding to fully replace myelography and myelo-CT [5, 11, 12, 13, 39]. Diagnosis with current imaging still is inaccurate in at least 15% of cases. Our preliminary experience using a constructive interference in steady state MR sequence is promising (Fig. 2c). MRI of the shoulder is valuable in detecting early secondary joint deformities [19, 40].

Fig. 2 a Cervical myelography: V-shaped anterior and posterior roots (*black arrow*) "pseudomeningocele" with root shadow of lesioned root (*white arrow*). **b** Myelo-CT: root-shadows of anterior and posterior roots (*arrows*) pseudomeningcele without recognizable root structure (*star*). **c** Constructive interference in steady state MRI with high resolution showing anterior rootlets (*arrow*) with computerized image magnification

Neurophysiological examination

Some find the results of preoperative neurophysiological studies overly optimistic [14, 18, 35]. Others combine evidence of nerve action potential and electromyography studies and are able to differentiate favourable conditions such as prolonged conduction block related to neuropraxia from more severe such as ruptures or avulsions as a basis for treatment decisions [2].

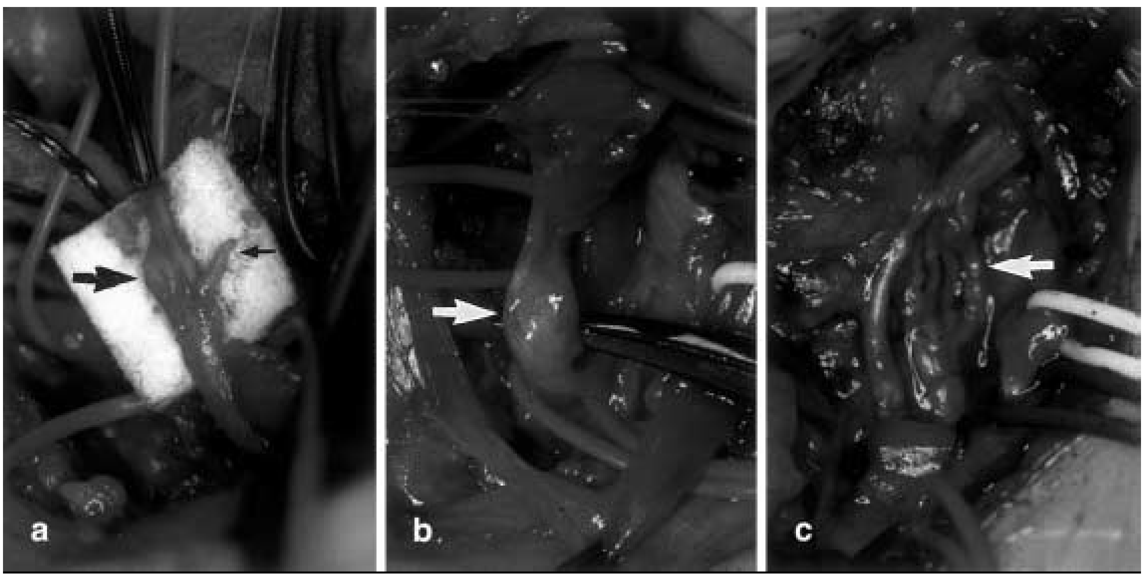
Operation

Surgical management and prognosis depend on accurate diagnosis of root avulsions to avoid ineffective repair. The plexus is approached through a transverse supra-lavicular skin incision. In upper plexus lesions (groups 1 and 2) the proximal roots C5–C7, the superior and middle trunk, and their divisions are exposed. In (sub)total lesions (groups 3 and 4) the lower roots and trunk are also exposed. Avulsion is evident if a dorsal root ganglion is seen (Fig. 3a) or a neuroforamen is empty. If a root seems in place, it may appear atrophic and pale although actually avulsed. Sensory evoked potentials recorded from spinal nerves should not show cortical responses when dorsal roots are avulsed [5, 20, 30]. Transcranial motor evoked potentials may contribute to assessing the functional status of anterior roots although practical use is still limited [37].

Particular problems arise when partial root avulsion is suspected from MRI, myelo-CT, or pre- and perioperative neurophysiological studies. The quality of a partially avulsed root, however, remains unknown, and therefore its suitability for neurotization. The myelin amount assessed intraoperatively in frozen sections is related to the quality and regenerative capacity of a proximal nerve stump as a source of outgrowing axons and may support decisions for repair [25]. Most other lesions are neuromas at truncal levels which extend to the foramen and distally into the various plexus divisions (Fig. 3b). A change in caliber may indicate a rupture. There is generally some continuity in a neuroma, as expressed by distal muscle response after a proximal stimulus; however, this is not of decisive relevance [2, 8].

Plexus reconstruction is individually tailored to the extent of the exposed lesion combined with clinical and other investigative assessments. The aim is to reinnervate the complete plexus combining grafts with transfers. Neuromas, regardless of whether in continuity, are resected and not neurolysed [2, 8]. The quality of proximal and distal stumps is assessed under the operating microscope and in frozen sections. Sural nerves, either resected by assisted endoscopy or under direct vision, are preferred for conventional grafting (Fig. 3c) [31]. To reconstruct completely or partially avulsed roots direct neurotisation or grafts to viable other proximal stumps are used. Transfers from extraplexial nerves as the accessory or intercostal nerves to the suprascapular or musculocutaneous nerve or to other distal nerve stumps are effective alternatives [2, 18, 26, 35]. As a last resource, the hypoglossal nerve should be reserved for

Fig. 3 a Avulsed spinal nerve root with dorsal ganglion (*large arrow*) and anterior motor root (*small arrow*). **b** Neuroma in upper trunk (*arrow*). **c** Plexus repair with sural nerve grafts (*arrow*)



direct motor neurotization. Partial denervation of the tongue may interfere with sucking and speech development (A.C.J. Slooff, personal communication). Reinnervation of the hand, i.e., the lower trunk, is of first priority in decision making regarding infants [2, 14, 15, 35]; elbow and shoulder function come next. The head and limb are immobilized in a bycast plaster for 3 weeks.

Outcome of microsurgical plexus reconstruction

Comparing results between different series is troubled by the use of different systems for assessment, as noted above. Based on series with a follow-up of at least 2.5 years and using the outcome system of Gilbert and Raimondi [2, 7], it can be concluded that in C5 and C6 lesions (group 1) over 80% of patients reach good shoulder function (stage 4 or 5, maximum 5), and results are even better for elbow function. In ruptures of C5, C6, or C7 (group 2 lesions) at least 65% reach good shoulder function (stage 4 or 5), 80% good elbow function (stage 4 or 5), and about 70% good wrist extension. In total lesions the chief aim is to restore hand function. Eventual results show good shoulder function (stage 5) in about 39% and useful elbow (stage 3) in 71%. Regarding hand function, 25% reach stage 4, 50% stage 3, and 20% stage 2. In children meeting the criteria for surgery results after plexus reconstruction are better than the natural history of such lesions [2, 7, 15, 16, 35, 40].

Rehabilitation

Despite plexus reconstruction temporary or definite neurological deficit may cause secondary contractures and deformities with negative functional implications. Most common is the medial rotation contracture at the shoulder, inflicting passive and active exorotation and abduction [2, 18, 21, 40]. The flexion and pronation deformity at the elbow is often seen in combination with the former and may be secondary to it [2]. To prevent these deformities as much as possible, children should receive an extensive rehabilitation program. Age-related skills form the starting points for treatment. In addition to long-term physiotherapy, children receive occupational therapy from the age of 1.5 year aimed at improving interaction of the arm in two-handed play and, at later stages, in daily self-care and school activities. Ortheses sometimes assist in preventing contractures or improving function. As muscle function may improve for years, treatment should continue under systematic supervision of a physiatrist.

Secondary surgery

Procedures are of two types: those to palliate effects of contractures and deformities and those to improve active function. The common medial rotation contracture causes glenoid deformities, posterior subluxation of the humeral head, and posterior capsular abnormalities, as demonstrated by MRI [19]. Orthopedic surgery is indicated in progressive deformities for correction or palliation, sometimes as early as the fourth month [2, 21, 40]. Musculotendinous transfers or osteteotomies may improve function but are seldom performed before the age of 4 years. In cases of widespread muscle weakness results are uncertain [2, 18, 40].

Team approach

Multidisciplinary aspects of treatment require formation of plexus teams in specialized centers. The management of children with OBPL requires the cooperation of a pediatric neurosurgeon, physiatrist, orthopedic and plastic surgeon, neurophysiologist, physiotherapist, and occupational therapist.

Future prospects

Prevention

Improved prenatal diagnostics (ultrasound) enables the selection of high-risk deliveries with large, heavy babies or breech positions in which expanded criteria for cesarian sections should be considered. Education of midwives, medical students, general practitioners, pediatricians, and physiotherapists, may either help to prevent OBPL or at least effect early referral of serious cases to specialized centers.

Additional investigations

Improved MR techniques will enable more exact imaging of small nerve structures such as spinal rootlets (Fig. 2c) and peripheral lesions at the fascicular level. Combined with perioperative histopathological examination of nerve stumps and possibly refined neurophysiological equipment, these will be of great importance in improving surgical management.

Although neurophysiological tests must be used in practical settings, further research is indispensable for improving our knowledge about functional regeneration of nerves and muscle and central nervous system changes in reaction to peripheral nerve lesions and their repair [3, 20, 22, 23, 26, 32]. Movement studies with electromyography registration may assist in understanding the development of certain abnormal movement patterns that are frequently observed. It is well recognized that the brain exhibits striking plasticity, especially during development. Metabolic imaging with positron emission tomography may allow detection of brain reorganization in relation to outcome after OBPL [6].

Treatment

A multidisciplinary team approach in not sufficiently recovering children should be standard for specialized centers. Future standardization of clinical and additional investigative assessment for each specialty involved will enable better multicenter comparison of treatment and outcome. Current microsurgical nerve repair seems to have reached its technical limits. Direct re-implantation or grafting of avulsed spinal roots to the spinal cord in primates or humans requires laminotomy and facetectomy and the manipulation of the spinal cord. This may cause spinal instability and cord injury [4]. Results do not yet justify the development of surgical procedures in infants. Moreover, it is not known whether the regenerative capacity of the root entry zone of the spinal cord is comparable to that of the peripheral nerves or to central nervous tissue. In the central nervous system axonal out growth is limited by many factors [10]. Research is focused on developing neutralizing agents to promote outgrowth of axons in central nervous tissue [10]. Donor site morbidity can be reduced further by the use of minimally invasive techniques [31]. Nerve repair by synthetic nerve guidance conduits is a future prospect [24, 36]. Nerve regeneration may be manipulated by the use of agents promoting nerve growth such as insulin-like growth factor 1 [29].

Conclusion

The outlook for children suffering from OBPL to regain the function of arms that are destined to be severely disabled if left untreated has improved thanks to the pioneering work of dedicated physicians in the twentieth century. Today integrated multidisciplinary treatment of OBPL has become a developing field of interest with challenging prospects for times to come.

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CHAPTER 3

PREOPERATIVE INVESTIGATIONS IN OBSTETRIC BRACHIAL PLEXUS PALSY

Based on:

Seminars in Plastic Surgery 2005; 19 :17-23

Preoperative Investigations in Obstetric Brachial Plexus Palsy

ABSTRACT

Treatment strategy in infants with obstetric brachial plexus palsy (OBPP) largely depends on clinical neurological examination and the degree of improvement in the first 3 months. Usually, less severe lesions show early recovery within a few months, and prognosis for spontaneous recovery is good. However, sometimes the time course of improvement may differ and will not allow clinical differentiation from more severe types of lesions with less favourable prognosis. Ancillary preoperative investigations assist in determining the type, level, and extent of the nerve lesion. The role of electrodiagnosis in preoperative assessment of OBPP is depreciated by many, whereas others stress the importance of combining results from electromyography and nerve action potential recording to discriminate between a nerve conduction block and a root avulsion or to predict the severity of axonal injury or degeneration. There is no role for motor-evoked potentials in OBPP yet. For imaging of the brachial plexus in infants, magnetic resonance imaging has surpassed computed tomography–myelography as modality of choice. High strength magnetic resonance scanners, applying different techniques in a non-invasive way allow imaging of plexus structures with great detail. Detection of different nerve lesion types is possible, such as root avulsions or nerve ruptures, formation of pseudomeningoceles, neuromas, or scarring as well as deformities of the shoulder joints. Magnetic resonance imaging is becoming a great aid as a preoperative investigation in determining treatment strategy in infants with severe OBPP.

Obstetric brachial plexus palsy (OBPP) is caused by a stretch injury to the brachial plexus. Reliable studies concerning prognosis are still lacking and, as a consequence, so are those concerning treatment. Most (75%–90%) infants will recover spontaneously, but babies with severe lesions will need microsurgical reconstruction of their damaged brachial plexus. Although the timing of and criteria for surgery are still a matter of debate, some consensus has been reached.[1–5] Preoperative ancillary investigations ideally should provide information on the type, extent, and location of the nerve injury and should be reliable indicators of prognosis. In OBPP neuro-physiological and imaging studies help to delineate the lesions and in many cases allow for an accurate diagnosis. When children meet the more or less accepted criteria for surgery, preoperative neurophysiological and imaging studies are performed, usually around the fourth month of age.[4,5] However, especially in babies and small infants, these investigations have their limitations.

The brachial plexus supplies sensory and motor innervation to the upper limb. It forms from the ventral rami of the C5–T1 spinal nerves. These nerves coalesce into three trunks along the posterolateral margin of the anterior scalene muscle. The upper trunk is derived from C5 and C6, the middle trunk from C7, and the inferior trunk from C8 and T1. Toward the periphery, the trunks divide into six divisions, finally coalescing into three cords, which are named the medial, lateral, and posterior cords.

There are four basic types of nerve injury.[6] Neurapraxia is a temporary conduction block. In axonotmesis the axon is severed but the surrounding nonneural elements are intact. In both types of injuries, spontaneous recovery is very likely. The other two, much more severe, types of nerve injury are usually managed by surgery at some stage. Neurotmesis is a complete postganglionic disruption of a nerve, and avulsion is a preganglionic disconnection of a spinal nerve from the spinal cord. In OBPP, most lesions are at the level of the supraclavicular plexus structures (i.e., proximal to the cords).

In OBPP in general there is still a lack of reliable indicators of prognosis on which treatment strategy can be based. As the success of nerve-grafting procedures in OBPP depends on the presence of regenerating axons in the proximal nerve stumps, the central diagnostic problem in severe OBPP for which a decision to perform surgery must be made is discriminating between neurotmesis and avulsion and especially reliably demonstrating spinal root avulsions in preoperative investigations.[7,8] When spinal roots are avulsed from the spinal cord, regenerative capacity is lost, as there is no connection with the central cell body. During surgery, a root avulsion is evident when the spinal ganglion is retracted extraforaminally (Fig. 4C). However, if an actually avulsed nerve root is not retracted, it may seem deceivingly normal at its exit from the foramen. Repair from such a nerve is doomed to be ineffective because there are no regenerating axons. The same holds true for intradural and intraforaminal ruptures of spinal nerve roots. With the demonstration of other types of lesions in preoperative ancillary examinations in OBPP peripheral to the foramina, it may be possible in the future to relate these to indications for surgery or to outcome. As yet it seems of less clinical importance, because most of the severe lesions necessitating surgery can readily be recognized and analyzed at operative exploration of the brachial plexus. An exception may be the electrodiagnosis of a prolonged nerve conduction block in which late spontaneous recovery can be expected.[9]

Clinical neurological examination in OBPP is the base for further treatment decisions. Ancillary investigations as yet delineate different types of nerve lesions and support treatment strategies. In the following pages, preoperative electrodiagnosis and different imaging techniques will be discussed.

PREOPERATIVE NEUROPHYSIOLOGICAL EXAMINATION

Electromyography

There are serious doubts about the value of electromyographic (EMG) studies in babies with OBPP, as a severe clinical picture is often in contradiction with optimistic EMG findings.[3,10–14] The EMG in babies with OBPP differs from adults in that denervation occurs and disappears much earlier. It can be found already on the fourth day and may have disappeared at 4 months. The very early denervation, as a sign of axonal degeneration, can be explained in infants by short distances and small diameter of the nerves. As compared with nerve size in adults, a 7.5–10 times faster denervation can be expected in infants. For adults, a 10–14-day period is normal, whereas a period of 1–2 days should not be too surprising in babies. Denervation activity usually disappears between day 10 and day 60 of age.[15] In many centers, EMG studies are performed at age 3–4 months.[16] By then, even children with completely avulsed C5 and C6 roots and a paralytic biceps muscle show a normal EMG on reflex-activated contraction of the biceps muscle. For this discrepancy between lack of functional muscle activity and EMG findings with motor unit potentials (MUPs), van Dijk and Vredeveld offer several explanations.[13,14,17]

An inadequate clinical examination may fail to register a slight muscle contraction. Because muscle fibers and corresponding motor units are 11-fold smaller in infants than in adults, the number of active MUs in infants is easily overestimated if the same-size EMG needle is used for both adults and infants.

Another explanation is the concept of luxury innervation. At birth there may still be a polyneural innervation in which nerves from multiple segments supply extra innervation to muscles. This pattern is reported to disappear and to be replaced by mononeural innervation from week 16 to 25 of gestation, up to 3 months of age.[18,19] In the absence of the original dominant innervation, as in root avulsions, this pattern probably persists, explaining the overly optimistic, near normal EMG findings. In contrast, and at the same time, the functional discrepancy with severe paretic muscles is striking. The reason for this may be that the central motor pathways do not primarily project to the anterior horn cells of these luxury nerves.

During infancy, central motor programs for different movement patterns are developed, the quality of which is dependent on afferent impulses. In OBPP, serious sensory deafferentiation interferes with the development of motor programs, resulting in inadequate, abnormal movement patterns and MUPs without effective movement, as is frequently observed.

If misdirection of axonal outgrowth occurs after a nerve lesion, different muscles — agonists as well as antagonists — may receive reinnervation from the same nerve, which in turn may lead to abnormal non-functional movement patterns, such as cocontractions, but MUPs in the EMG are readily registered.

In the EMG of infants with OBPP, the recording of MUPs is not a reflection of functional muscle activity based on a developing central motor program but merely an indication of some axonal continuity from the spinal cord to the muscle.

Nerve Action Potentials

Despite the restrictions of EMG in infants, according to some investigators, the combination of EMG with recording nerve action potentials from the median and ulnar nerves at the elbow after stimulation at the wrist may significantly add to the clinical assessment in determining the nature and level of a lesion. In individual cases, it is thus possible to predict the prognosis and the need for surgery.[7,20] With this technique, a nerve conduction block or avulsion can be revealed.[7,20] If a nerve action potential is comparable to the healthy limb, a lesion is not degenerative, but there is either a conduction block or avulsion in which the sensory ganglion has at least remained intact. In avulsions, the EMG usually shows signs of denervation. If denervation activity is lacking in the EMG, then the lesion is a prolonged conduction block. In such a case, surgery should be deferred because spontaneous recovery is to be expected. In combining EMG and nerve action potential recordings, some investigators have even been capable of distinguishing further between axonotmesis and neurotmesis.[7,20]

Somatosensory Evoked Potentials

After electric stimulation of a peripheral sensory (e.g.,the digital) nerve, somatosensory evoked potentials (SEPs) are recorded over the somatosensory cortex. A normal cortical response in OBPP indicates that a fair amount of sensory fibers in the brachial plexus and in the dorsal roots are conducting normally. However, it lacks qualitative, quantitative, and localization precision regarding the type and extent of a lesion. Direct root SEPs during surgery are more informative, but they nevertheless have comparable restrictions and do not offer information on the condition of the ventral, motor roots.[17,20]

Motor Evoked Potentials

Preoperative adequate evaluation of the functional integrity of anterior cervical spinal nerve roots by motor evoked potentials (MEPs) in OBPP is not possible, as direct root registration is ‘‘not done.’’ Transcranial electrical MEPs with the usual registration of muscle activity by EMG is not diagnostic for single or multiple anterior root lesions because most muscles have a polyneural-innervation.[13,14,20] Only intraoperative MEP and direct registration at the nerve roots offer information on the proximal continuity of anterior spinal roots, but they also do not reflect the quality and recovery potential of an individual nerve root.[21] The technique is not reported in babies with OBPP. Because of the relative lack of myelin in the central white matter in infants compared with adults, it is presumed that cortical depolarization by transcranial electrical stimulation is more difficult to achieve. Therefore, MEPs have not been introduced as a preoperative or intraoperative diagnostic tool in babies with OBPP.

PREOPERATIVE IMAGING

In infants with OBPP who have severe enough lesions to meet the more or less accepted criteria for performing surgery, imaging is part of the preoperative analysis and is usually performed in or around the fourth month.[1–3] As mentioned before, the key issue in severe OBPP is to reliably exclude or demonstrate preganglionic spinal nerve root lesions and, even more demanding, to show precisely the combinations of intact or avulsed anterior and dorsal roots as well as complete or incomplete nerve root avulsions. Hemilaminectomy and intradural inspection of root entry zones of relevant spinal cord levels offer the most reliable control for root avulsions but are only feasible in adults.[22,23] Even with direct inspection and combined intraoperative, direct root SEP recordings, some uncertainty remains regarding the functional integrity or recovery potential of anatomically intact roots. In babies, such operative procedures are not performed, which is why a golden standard to assess the accuracy of any preoperative diagnostic study remains elusive.

Conventional X-Ray

As many associated lesions are described in OBPP, X-ray studies are of value in the newborn to show fractures or luxations of the cervical spine, humerus, or clavicle. Additional radiographs of the shoulder are of interest in a long-term follow-up scheme to assess osseous deformities of the glenoid fossa and humeral head.[24]

A chest X-ray, or ultrasound as an alternative, should be a standard procedure for the diagnosis of a diaphragmatic paralysis caused by a C4 or phrenic nerve lesion. To prevent postoperative respiratory problems, especially during bottle or breast feeding, plication of a paralytic diaphragm is recommended before exploring the brachial plexus and is even more important when intercostal nerve transfers are considered.[25]

Computed Tomography–Myelography

In the past decade, computed tomography (CT)–myelography was the preferred diagnostic tool in OBPP in most centers. Many investigators experienced a higher reliability in the assessment of intradural root avulsions than with magnetic resonance imaging (MRI) techniques.[22,25–29] Disadvantages of CT-myelography, however, include the need for anesthesia, intrathecal contrast application, and radiation. Several shortcomings of CT myelography are also well known.[22,23,28,29] Although contrast-filled pseudomeningoceles (Fig. 1), subarachnoid space deformity, or missing root shadows in CT myelography are associated with root avulsions, they are known to show false-positive or false-negative results. Because there is a discrepancy between the root exit zone at the spinal cord and the corresponding intervertebral exit foramen, uncertainty about the determination of the correct spinal level may prevail even when using digital reformation techniques based on 1.5-mm transverse slices. An avulsion can only be ruled out when relevant anterior and posterior root shadows can be traced from the spinal cord to the exit foramina.[25,29] In their intra and extraforaminal course, CT-myelography fails to demonstrate ruptures or other types of lesions of spinal nerves.

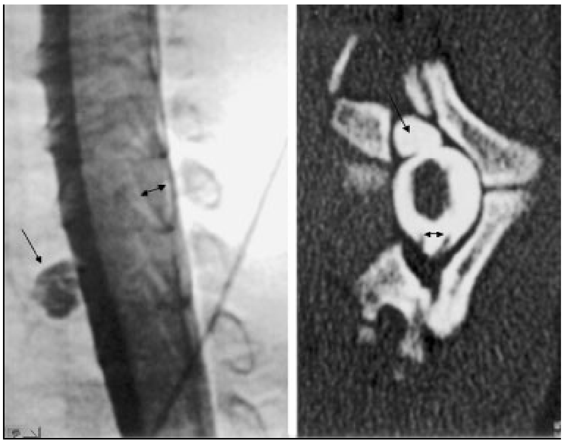

Figure 1 Computed tomography–myelography in an infant of 4 months with obstetric brachial plexus palsy. (**A**) Myelogram; in a contrast-filled pseudomeningocele (arrow), root shadows are demonstrated. Intact anterior and posterior roots (double arrow) on the non involved side. (**B**) Computer tomography–myelography; no recognizable root shadows in the contrast-filled pseudomeningocele (arrow). Intact roots on the non involved side (double arrow)

Magnetic Resonance Imaging

MRI has become the preferred examination in many centers for imaging the brachial plexus in infants because of its non-invasive character.[27,30] High-strength (at least 1.5 Tesla) MR machines create high-resolution images that enable visualization of different types of nerve lesions. Detection of nerve root avulsions and intraspinal nerve lesions is most valuable for treatment strategy, as mentioned. For imaging of intradural spinal nerve segments, three-dimensional (3D) constructive interference in steady-state MRI provides excellent-quality imaging.[16,25] This is a heavy-weighted T2 sequence with a strong and constant signal for cerebrospinal fluid. With 3D fast spin echo T2 MR imaging with gray-scale inversion, fine-quality imaging can also be achieved.[27] Studies of babies can be performed under mild sedation. Different MR techniques ideally even allow the demonstration of other types of lesions in the extraforaminal, peripheral brachial plexus nerves, such as scarring, edema, or formation of neuromas.[30,31]

Secondary deformities of the shoulder in OBPP can also be demonstrated with MR and

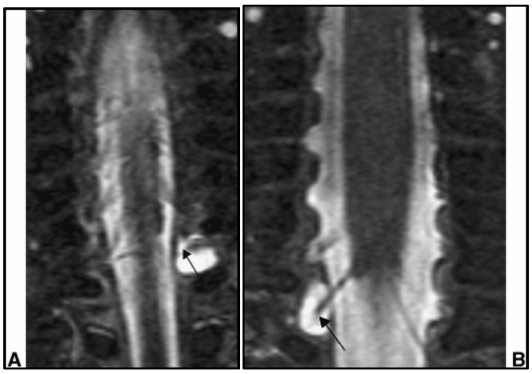

Figure 2
Three-dimensional constructive interference in steady-state magnetic resonance imaging, coronal images in an infant of 4 months with obstetric brachial plexus palsy. (A) Interrupted anterior root shadow in a pseudomeningocele. (B) Pseudo-meningocele in which the anterior root can be traced to the spinal cord.

are of interest in follow-up schemes. However, around the fourth month of age, these deformities rarely have direct orthopaedic surgical consequences and thus have limited value as preoperative diagnostics.[32,33]
MR studies avoid radiation, are non-invasive, can be performed under mild sedation in most infants, are less time consuming than CT-myelography studies, do not put a claim on anesthesia personnel, and are thus more cost effective.

INTRASPINAL IMAGING; NERVE ROOT AVULSIONS

Avulsions can be complete, affecting both anterior and posterior roots, or incomplete, with selective avulsion of either anterior or posterior roots or even of part of their contributing rootlets. Avulsions are often associated with pseudomeningoceles, which occur after disruption of nerve root sleeves, thus allowing cerebrospinal fluid to extrude from the subarachnoid space (Fig. 2A). Pseudo-meningoceles can be isolated injuries with intact or at least not avulsed nerve roots (Fig. 2B). When root shadows of spinal nerves can be traced from the spinal cord to the respective exit foramina, there is no avulsion. [22,25,29,34]
If there is an interruption of a root shadow on an axial image, the coronal image should be checked for confirmation, because continuity is easily missed in the axial 2-mm slices. Avulsion is likely when images in both planes fail to demonstrate continuous roots. Demonstration of partial disruption of rootlets or avulsion of either an anterior or posterior root is incidentally possible. Sometimes recoil of a root, showing a blunt stump at the spinal cord, is suspected after preganglionic disruption in the absence of a pseudo-meningocele (Fig. 3) In pseudomeningoceles, which are isointense to cerebrospinal fluid, delineation and tracing of root shadows can be difficult. Intact, avulsed, compressed, or scarred intradural spinal nerves are possible.

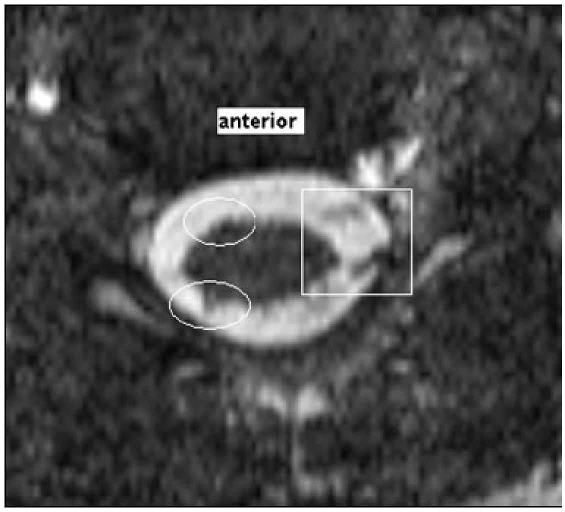

Figure 3 Three-dimensional constructive interference in steady-state magnetic resonance imaging, axial view at level C7. Intact roots on noninvolved side (square) and recoil of avulsed anterior and posterior roots on involved side (ovals).

EXTRASPINAL BRACHIAL PLEXUS IMAGING

There are no specified reports on imaging of spinal nerve lesions from the dural exit and along their intervertebral intraforaminal course. Roots can be traced to their exit foramina on axial and coronal views, using 3D constructive interference in steady-state MRI or 3D fast spin echo T2, but a study on differentiation and correlation of lesion type is still lacking, as

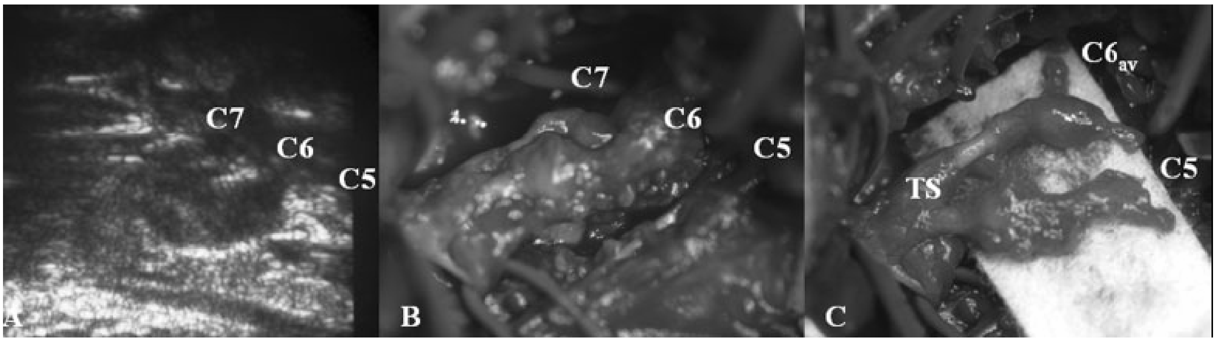

Figure 4 Preoperative ultrasonography (ATL, 7.5-MHz transducer, Bothell, WA) of left supraclavicular region and comparable operative views in an infant of 5 months with obstetric brachial plexus palsy. (A) The plexus structures are hypointense on ultrasonography and can be traced from the foramina C5, C6, and C7 toward the periphery (from right to left). The proximal part of C5 is not demonstrated. From the level of C6 to the superior trunk a thickened irregular mass is shown. (B) Intraoperative view of supraclavicular plexus comparable to the ultrasonography image. From C6 to the superior trunk fibrosis and neuromatose thickening. The proximal intact root C5 (not shown) was turned over into the scalenus muscle and dissected free. (C) After dissection of the neuroma, the lesion proved to be an avulsion of C6 (C6av) and a neurotmesis of C5. The spinal ganglion of C6 and the preganglionic ruptured anterior and posterior roots are clearly demonstrated.

intraoperative control in this trajectory in babies (and even adults) is troublesome, requiring more or less extensive foraminotomies. Some conclusive, though not in all cases, additional evidence on continuity of a spinal nerve may be gained by intraoperative monitoring techniques (SEP and MEP), which have their own restrictions and limitations, as mentioned before.

IMAGING OF THE EXTRAFORAMINAL SPINAL NERVES AND BRACHIAL PLEXUS

Imaging of roots, trunk divisions, and cords is possible using high-strength MRI scans with T1- and T2- weighted different imaging techniques in axial, coronal, and sagittal planes individually adapted and planned parallel to the angle of the brachial plexus with additional use of reformatting techniques.[30,31,35]
Precise imaging of the extraforaminal brachial plexus requires scanning times of more than 1 hour, posing problems to maintaining protocols with only sedation, thus requiring anesthesia anyway. Most reports describe posttraumatic plexus injuries in older children and adults. Birchansky elegantly describes MR imaging in babies.[30] Normal or even pathologic nerves are isodense to adjacent muscle but hypodense to surrounding fat. On T2-weighted and short tau inversion recovery images, the plexus is slightly hyperintense to muscle. Posttraumatic sequelae such as neuromas or fibrosis mainly affect the configuration of plexial nerves but do not usually alter signal intensity in T1 or T2 imaging. Some increased signal intensity in T2-weighted images may be observed. After complete or incomplete nerve ruptures, neuromas are formed. In a neuroma, the normal fascicular anatomy is lost and will be replaced by a thickened mass of disorganized proliferating axons and fibrous tissue, which can be recognized using MRI as a fusiform mass. Depending on the severity of the OBPP, the localization and number of nerve ruptures and consequent formation of neuromas may vary. Posttraumatic perineural fibrosis of the brachial plexus can be focal or diffuse. It may be recognized as thickening of plexial structures with ragged borders.[30]
Nevertheless, exact discrimination of all possible combinations of nerve lesions in OBPP is not possible with MRI. Although at present, demonstration of neuromas or fibrosis of the brachial plexus in infants with insufficient recovery of function would not generally influence treatment strategy, the more and more precise imaging quality adds to the understanding of different plexus lesions and may in the future have a growing effect on treatment decisions.

PERIPHERAL MR NEUROGRAPHY

In adults, pathology of peripheral nerves can be studied by MR neurography - a technique enabling demonstration of nerve fascicles in cross sections of larger nerves in adults.[31] As MR neurography is not suited to study smaller nerves, even those of sizes comparable with the trunks or cords in babies, it cannot yet be applied in infants with OBPP.

ULTRASONOGRAPHY

The newest generation of ultrasonography equipment allows imaging of supraclavicular plexus structures in infants. In individual cases, it is possible to demonstrate different pathology; for instance, neuromas or missing nerve roots. To check the accuracy of this imaging technique, preoperative images should be compared with intraoperative findings (Fig. 4). With only preliminary experience, valuation of the method is premature, and as yet, studies are not reported on this issue in OBPP.

CONCLUSION

In the developing field of treatment of OBPP, skilful and creative combined use of electrodiagnosis and imaging techniques not only allow characterization and differentiation of nerve lesions with ever-increasing precision but also may provide growing insight into largely unexplored central mechanisms of functional plasticity.

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CHAPTER 4

DETECTION OF ROOT AVULSION IN THE DOMINANT C-7 OBSTETRIC BRACHIAL PLEXUS LESION. EXPERIENCE WITH 3D-CISS MRI AND ELECTROPHYSIOLOGY

Based on:

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DETECTION OF ROOT AVULSION IN THE DOMINANT C7 OBSTETRIC BRACHIAL PLEXUS LESION: EXPERIENCE WITH THREE-DIMENSIONAL CONSTRUCTIVE INTERFERENCE IN STEADY-STATE MAGNETIC RESONANCE IMAGING AND ELECTROPHYSIOLOGY

OBJECTIVE:
Preoperative, reliable detection by ancillary investigations of spinal nerve root avulsions in infants with severe obstetric brachial plexus lesions to avoid ineffective operative repair from deceivingly intact but actually avulsed nerve roots.

METHODS:
Ten infants were selected with an infrequent, severe dominant C7 lesion, primarily because of the anatomically distinct supraclavicular course of this spinal nerve. Three-dimensional constructive interference in steady-state magnetic resonance imaging (3D CISS MRI) studies under mild sedation were performed and evaluated for detection of avulsed nerve roots by two experienced neuroradiologists. Preoperative electrodiagnostics (electromyography and somatosensory evoked potentials) as well as intraoperative somatosensory potentials and muscle contractions after electrostimulation were recorded. Preoperative and intraoperative ancillary investigations were correlated with intraoperative findings in eight patients and clinical status in two children who recovered spontaneously.

RESULTS:
Despite two minor motion artifacts, the quality of the 3D CISS MRI studies was good. In 8 of 10 patients, prediction of root continuity was consistent with operative or clinical findings, and 2 remained doubtful. Preoperative and intraoperative electrodiagnostics tended not to correlate with intraoperative findings in this small, selected group.

CONCLUSION:
3D CISS MRI provides good images of anterior and posterior spinal roots in infants with obstetric brachial plexus lesions. Images seem to allow accurate prediction of root avulsion in the majority of patients. In this study, electrodiagnostics were of limited value.

The success of nerve grafting procedures in obstetric brachial plexus lesions (OBPLs) that require surgical reconstruction depends on the presence of regenerating axons in proximal nerve stumps (1, 25). When roots are avulsed from the spinal cord, regenerative capacity is lost, because there is no connection with the central cell body. Root avulsions, however, can be very difficult to identify. If an empty neuroforamen is encountered or when a spinal ganglion is retracted extraforaminally, avulsion is evident. If an avulsed nerve root is not retracted, it may seem deceivingly normal, but because it lacks regenerating axons, repair from such nerve stumps is ineffective. Hemilaminectomy and intradural inspection of root entry zones at relevant spinal cord levels offers the most reliable control of root avulsion (3, 14), but although practiced in adults with traumatic lesions, this is not really feasible in infants (15). Therefore, additional imaging and electrophysiological studies focus on the issue of how to reliably detect complete or partial preganglionic nerve root lesions. Even when imaging is combined with intraoperative neuromonitoring, diagnosis of root avulsions in OBPLs remains inconclusive in at least 15% (6, 14). Computed tomographic (CT) myelography and routine two-dimensional magnetic resonance imaging (MRI) techniques still lack resolution in infants, whereas other techniques are technically too demanding (2, 4, 5, 7, 11, 18, 21, 24).

A new, fully flow-compensated, three-dimensional (3D) MRI technique with T2 weighting was introduced, the so-called 3D constructive interference in a steady-state (3D CISS) technique. We report the use of this 3D CISS MRI technique since 1998 and additional electrophysiological studies in 10 patients with a dominant C7 lesion (12, 17, 22) (*Table 1*) to detect root avulsion.

PATIENTS AND METHODS

Patients

From 1995 to 2003, 490 infants with OBPLs presented at the Vrije Universiteit University Medical Center in Amsterdam, the Netherlands. From this group, 14 infants (5 boys and 9 girls) presented with a severe dominant C7 lesion, which is not an isolated or exclusive C7 lesion but rather a subtotal plexus lesion with dominant paralysis of C7 innervated muscles according to the Narakas classification (*Table 1*; *Fig. 1A*) (12). Four children examined before 1998 only by CT myelography were excluded; the remaining 10 children were the subjects of this study. In OBPLs in general, ancillary investigations, as mentioned below, are scheduled, and surgery is indicated between Months 4 and 6 if there is either failure to recover biceps function, whether or not in combination with failure to recover extension of the elbow, wrist, fingers, and thumb within 4 months from birth, or evidence of a severe lesion with Horner's sign, persistent hypotonic paralysis of the arm, persistent phrenic nerve paralysis, persistent signs of denervation in electrophysiological studies, and suspicion of root avulsion on diagnostic imaging (1, 17, 22).

The same time schedule applies to the dominant C7 lesion in which failure to recover shoulder adduction and elbow extension, whether or not in combination with wrist extension, is predominant.

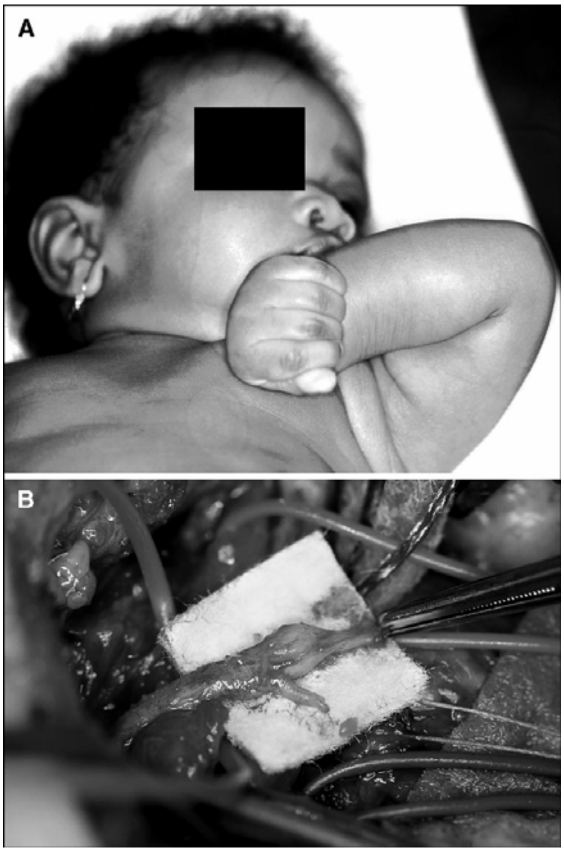

FIGURE 1. A*, photograph of infant with a dominant C7 lesion. Weakness of shoulder adductors, elbow, and forearm extensors.* **B,** *intraoperative photograph of spinal root avulsion with retracted ganglion proximally connected to sensory or posterior root (forceps) and just distal from the ganglion, the motor or anterior root.*

TABLE 1. Narakas classification of obstetric brachial plexus lesions

Classification obstetric brachial plexus lesions	Lesion	Clinical presentation
Group 1	C5, C6 or superior trunk	Paralysis of deltoid, shoulder exorotation, and biceps
Group 2	C5, C6 or superior trunk, C7 or medial trunk	Same as Group 1, with paralysis of extension of elbow, wrist, and digits. Typical "waiter tip" hand
Group 3	C5, C6 or superior trunk, C7 or medial trunk, incomplete C8, T1, or inferior trunk	Except for some finger flexion, virtually complete paralysis
Group 4	Complete plexus	Complete flaccid paralysis of upper extremity Horner sign
Group 4, dominant C7	C7 or medial trunk, incomplete other plexus structures	Paralysis of shoulder adduction and elbow extension and mostly in different degree of wrist and finger extension. Varying incomplete other muscle deficits.

Imaging: 3D-CISS MRI

In all patients, a 3D CISS MRI study (1.5 T; Siemens Magnetom; Siemens Medical Systems, Inc., Erlangen, Germany) was performed. Infants less than 4 months of age were sedated with chloral hydrate suppositories. Children older than 4 months were sedated with intramuscular injection of pethidine, droperidol, and chlorpromazine. During scans, all infants were controlled by video camera and monitoring of heart rate, breathing, and oxygen saturation. The MRI protocol contained scouts, sagittal T1-weighted spin echo, and coronal T2-weighted turbo spin echo for planning purposes and coronal as well as axial CISS studies. 3D-CISS is a heavily weighted T2 sequence with a strong and constant signal for cerebrospinal fluid. Two experienced neuroradiologists reviewed the studies by consensus. The quality of the studies was graded as *good* when resolution enabled clear discrimination of ventral and dorsal nerve roots in the intraspinal course at all cervical levels and as *sufficient* when resolution was just sufficient, despite slight (motion) artefacts, to allow interpretation of ventral and dorsal roots at levels C5–T1. If a study could not be interpreted at one or more cervical levels, it was qualified as *insufficient.* A spinal root was defined as avulsed when root shadows could not be identified on both axial and coronal images or could not be traced continuously from the spinal cord to the exit foramen, regardless of the presence of pseudomeningoceles. Predictions of root avulsions, intact intraspinal roots, or doubtful continuity and report of the affected cervical levels were correlated with operative findings.

Electrodiagnostics

Preoperative electromyographic (EMG) screening was performed with concentric needle electrodes in all patients. Several muscles were investigated, depending on the extent of muscle weakness. Signs of denervation looked for were fibrillation potentials at rest. During voluntary contraction, the shapes and amplitudes of motor unit potentials were identified if possible. In addition, an estimation was made of the maximal recruitment pattern. Because these studies were not performed according to a fixed protocol and because of the lack of cooperation of small infants, only the presence of fibrillation potentials as a sign of denervation on the needle EMG study at rest was used.

Preoperative somatosensory evoked potentials (SSEPs) from both median nerves were recorded with surface electrodes in the neck at the level of C7 and at the cranium over postcentral regions (reference at Fz). No sedation was administered. With a stimulus rate of 4 Hz, 2 - 500 responses were averaged. SSEPs were scored as *normal* when they were present and symmetrical and as *absent* when they were not reproducible. Those SSEPs that were more than 50% lower in amplitude compared with the contralateral side were scored as *low amplitude*, indicating axonal loss.

Intraoperative SSEPs were recorded in the eight patients operated on with surface electrodes from the level of C7 in the neck and the contralateral postcentral region, with a common reference at the forehead (Fz). Direct root stimulation was performed with bipolar hook electrodes using square impulses (1- to 10-mA constant current, 0.04-ms duration, 1.5 Hz).

Anesthetics that interfere with monitoring, such as inhalation anesthetics, were avoided, and only intravenous ketamine and propofol were used. No muscle relaxants were used. SSEPs from the contralateral median nerve were routinely performed as a control. After electrical stimulation of spinal nerves, motor reactions were observed, located, and palpated in the muscles of the affected extremity during operations and subsequently noted.

Operation

In all children operated on, the supraclavicular brachial plexus was exposed through an incision parallel to the clavicula. A root avulsion of C7 was evident when a spinal ganglion was recognized (*Fig.1B*) or verified by intra-operative histology. A neuroma in C7 or the medial trunk theoretically excluded a complete preganglionic lesion, because this is formed only when the regenerative capacity of the nerve is preserved. Proximal continuity was doubted when an intraforaminal, seemingly intact root was thin and pale and did not show a neuroma or spinal ganglion. Findings at other levels were recorded. Reconstructions were individually tailored accordingly.

Table 2. Operative findings, plexus reconstruction, and results of three dimensional constructive interference in steady state magnetic resonance imaging, electromyographic, and somatosensory potentials in Group 4 dominant C7 lesions (n=10)[a]

					3D CISS MRI				EMG denervation at rest			SSEP	
Pt.	C7 operative finding	C5/C6/Tr Sup	Age at operation (mo)	Reconstruction	Pseudocele	Root shadows coronal	Root shadows axial	Predicted avulsion C7	Triceps	Forearm extensor	Biceps	Preoperative median N SSEP	Intraoperative C7 SSEP C7 motor reaction
1	Avulsed, attached to superior trunk	neuroma	6	C5-C7 C5-posterior cord C6-lateral cord N XI-suprascap N	Yes/small	Absent	Inadequate	yes[b]	-	-	-	Low amplitude	Present Triceps
2	Avulsed	Normal	4	N XI- C7 motor C sens-C7 sens	Yes	Present, not continuous	Present, not continuous	Yes	+/-	+/-	-	Normal	Absent No
3	Avulsed	LIC	6	N XI-C7 motor C4 sens-C7 sens Tr Sup :neurolysis	No	Absent	Absent	Yes	-	-	-	Absent	Absent No
4	Avulsed, ganglion in foramen C7	LIC	7.5	N XI-C7 motor C4 sens-C7 sens	Yes	Absent	Absent	yes[b]	-	-	-	Absent	Absent Pectoral
5	Avulsed, proximal stumps ingrowth in C8	LIC/ normal	7	N XI- medial trunk (triceps reactive fascicle)	Yes	Absent	Absent	Yes	-	-	-	Normal	Present Triceps and pectoral
6	Thin C7, doubtful avulsion	Normal	7	N XI-medial trunk (triceps reactive fascicle)	Yes	Present, not continuous	Present, not continuous	Doubt	+	+	-	Normal	Present Triceps
7	No operation			Late spontaneous recovery	Yes	Absent	Absent	Yes	-	-	-	Absent	N.i.
8	Intact C7, normal C7 function, no active shoulder exorotation	Normal	13	Inspection C7 N XI- suprascap N	No	Present	Present	No	-	-	-	Normal	Present Triceps
9	No operation			Spontanous recovery	Yes	Ant present Post doubt	Ant present Post doubt	Partial	-	-	-	Normal	N.i.
10	Continuous root in foramen, doubtful avulsion	LIC	5	N XI – C7 (non-SEP reactive part)	Yes	Absent	Absent	yes	+	-	-	Low amplitude	Present No

[a] 3D CISS MRI, three-dimensional constructive interference in steady state magnetic resonance imaging; EMG, electromyography; SSEP, somatosensory evoked potentials; LIC, lesion in continuity; N XI, accessory nerve; Suprascap N, suprascapular nerve; N.i., not investigated; Sens, sensory; Tr Sup, superior trunk; ant, anterior; post, posterior; pt, patient; mo, months
[b] MRI studies of sufficient quality. EMG (-), no denervation; EMG (+), sole denervation; EMG (+/-), denervation in combination with reinnervation or voluntary activity.

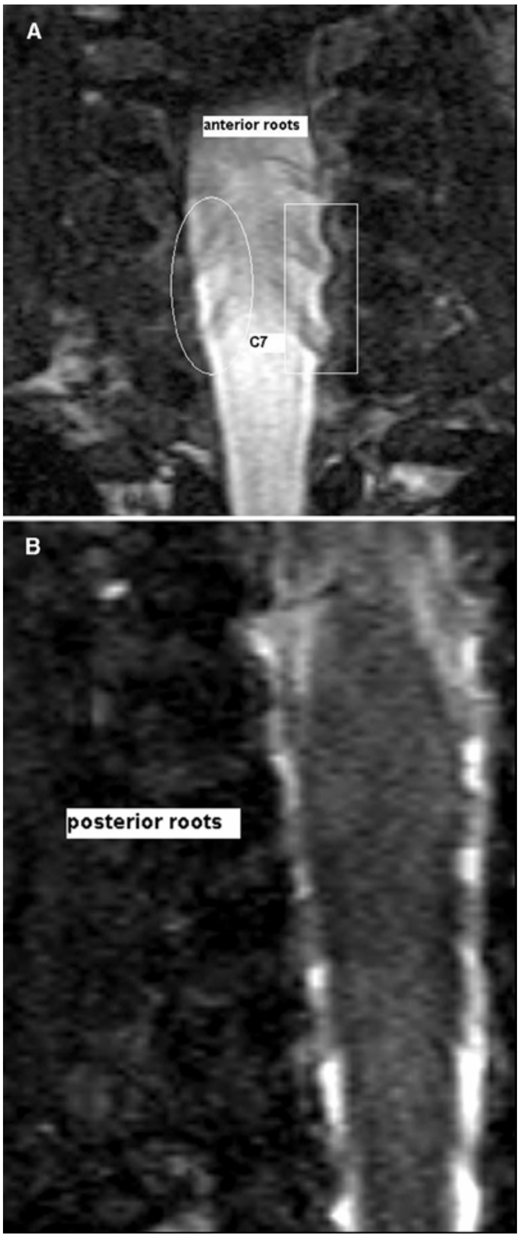

FIGURE 2. *Patient 8. 3D CISS MRI scan, coronal view at the level of* ***(A)*** *the anterior roots and* ***(B)*** *the posterior roots (*outlined areas*), showing continuity of roots and discrimination of rootlets in a child who recovered spontaneously with a dominant C7 lesion.*

FIGURE 3. *Patient 2.* **A**, *3D CISS MRI scan, axial view at spinal level of C7 roots. Pseudomeningocele with interrupted root shadows, in the case of a C7 root avulsion on the right side. Root shadows on the nonaffected side are clearly visible (*oval*).* **B**, *coronal view in the same patient showing the level of the anterior roots and a pseudomeningocele at C7 (*oval*), in which root shadows cannot be discriminated.* **C**, *coronal view at the level of the posterior roots. In the pseudomeningocele, a possible stump of the avulsed root can be discriminated in the pseudomeningocele (hypodense structure)*

RESULTS

Patients

In 10 patients with a dominant C7 lesion and insufficient spontaneous recovery, ancillary investigations were performed at a mean age of 5.5 months (range, 3.5–7 mo) (*Table 2*). Seven children were operated on for the typical C7 palsy at a mean age of 6.0 months (range, 4–7.5 mo). However, three patients showed slow spontaneous recovery, of which two were not operated on, whereas the third child underwent late plexus surgery at 13 months for lack of active shoulder exorotation not related to C7 pathology but with inspection of the C7 root.

Imaging

The quality of 10 3D-CISS MRI studies was good in 8 and sufficient, because of some motion artifacts, in 2 (*Table 2*, Patients 1 and 4). In the coronal images, the cerebrospinal fluid (CSF) signal is more homogeneous, probably because CSF flow occurs within the imaging slab, enabling more complete discrimination of root shadows over their entire intradural length. Examples showing the quality of the studies are presented in *Figures 2, 3,* and *4*.
Of the eight patients operated on, on the basis of the MRI, avulsions were predicted in six patients (*Table 2*, Patients 1–5 and 10), of whom five (Patients 1–5) (*Table 3*) were confirmed intraoperatively by demonstration of a dorsal root ganglion (*Fig. 1B*). Root continuity was predicted and confirmed in one (Patient 8). In two children, a predicted doubtful continuity Patient 6) and an avulsion (Patient 10) could not be confirmed, because doubt about the continuity of the C7 root remained even after intraoperative inspection. Assuming that the C7 roots were not avulsed in the two remaining children, who recovered spontaneously and thus were not operated on, then prediction of root continuity was right in one and wrong, because avulsion was predicted, in the other.

Electrodiagnostics

In preoperative EMG studies (*Tables 2* and *4*), denervation signs in the triceps muscle and/or extensors of the forearm were absent in the five patients in which avulsion of C7 was confirmed at operation. One infant (Patient 6) with doubtful avulsion on the MRI scan but a seemingly intact root intraoperatively showed complete denervation. The preoperative EMG study of the biceps muscle showed voluntary activity in all children. SSEPs from the ipsilateral median nerve were absent in three children (*Tables 2* and *4*). One eventually showed full spontaneous recovery; the other two had avulsions of C7 and lesions in continuity of the superior trunk.
Intraoperative direct SSEPs from C7 roots in the operative patients showed five positive recordings, even in two confirmed avulsions (Patients 1 and 5) (*Tables 2* and *5*). Direct electrical stimulation of C7 roots provoked motor reactions in the triceps and/or pectoral muscles in five children, even in three infants with avulsed C7 roots. Motor reaction was absent in two patients with proven avulsion of C7 (*Tables 2* and *5*).

Operative Findings

Clear avulsions with extraforaminally retracted spinal ganglia were confirmed at operation in five patients. Continuity was confirmed in one patient (Patient 8) but remained doubtful in two patients (Patients 6 and 10), in whom C7 roots seemed to be continuous within the foramen but were thin and pale. Two cases could not be verified because of conservative treatment. Roots at other levels did not clearly show preganglionic lesions. Depending on the clinical picture and the lesions found in other parts of the plexus, reconstructions were individually tailored (*Table 2*).

TABLE 3. Three-dimensional constructive interference in steady-state magnetic resonance imaging versus operative or presumed findings in 10 patients with a dominant C7 root lesion[a]

Operative finding: 3D CISS MRI	Avulsion of C7 spinal ganglion	Intact root C7		Doubtful continuity C7
		Operated on	Not operated on	
Predicted avulsion C7	5		1[b]	1
Predicted continuity C7		1	1[b]	
Doubtful continuity C7				1

[a] 3D CISS MRI, three-dimensional constructive interference in steady-state magnetic resonance imaging.
[b] Two patients showed spontaneous recovery and were not operated on but were presumed to have intact C7 roots.

TABLE 4. Preoperative denervation in triceps and/or extensors forearm, median nerve somatosensory evoked potentials versus operative findings in 10 obstetric brachial plexus lesion patients with a dominant C7 lesion[a]

Operative finding: neurophysiology	Avulsion of C7 (n = 5)	Intact root C7 (n = 1)	Doubtful continuity C7 (n = 2)	Spontaneous recovery (n.o.) (n = 2)	Total investigated
Denervation triceps and/or extensor digitorum	1	1	1	0	10
SSEP median nerve present	3	1	2	1[b]	10
SSEP median nerve absent	2			1[b]	

[a] SSEP, somatosensory evoked potentials.
[b] Two patients showed spontaneous recovery and were not operated on (n.o.).

TABLE 5. Intraoperative C7 root somatosensory evoked potentials and motor reactions versus operative findings in 8 obstetric brachial plexus lesion patients with a dominant C7 lesion[a]

Operative finding: SSEP/motor reaction	Avulsion of C7 (n = 5)	Intact root C7 (n = 1)	Doubtful continuity C7 (n = 2)
No SSEP	3		
Present SSEP	2	1	2
No SSEP and no motor reaction	2		
No SSEP and motor reaction of triceps and/or pectoral muscle	1		
SSEP and motor reaction	2	1	1
SSEP and no motor reaction			1
No SSEP and no motor reaction + MRI-predicted avulsion	1		

[a] SSEP, somatosensory evoked potentials; MRI, magnetic resonance imaging.

DISCUSSION

The application of the 3D CISS MRI technique in OBPLs provides high-quality diagnostic imaging in the preoperative diagnosis of root avulsions in infants as exemplified for the severe dominant C7 lesion. However, in the case of OBPLs, this new technique cannot be verified by direct intraoperative intradural visual confirmation of root avulsion. Therefore, it is still the practice to add electrophysiological assessment, which is presumed to predict preganglionic spinal root integrity more reliably. Although the described ancillary investigations are performed when indicated in all groups of OBPL patients, the interpretation of these investigations into cause and effect of a clinical presentation remains very complex, because it is hardly ever possible to deduce symptoms and pathological findings to just one root (12, 22). Anatomically, the C7 root is the only root not joined by another spinal nerve at the truncal level but runs a single supraclavicular course as the medial trunk to contribute distally primarily to the posterior division. Neuroma formation in this trajectory means preserved regenerative capacity of the damaged nerve and theoretically excludes a complete preganglionic lesion (25). The clinical picture of this subtotal plexus lesion is dominated by a paralysis of primarily C7 innervated muscles (shoulder adductors, triceps muscle, and forearm extensors) (*Fig. 1A*) associated with incomplete lesions and functional deficits of at least the neighboring roots C6 and C8, which, to different degrees, do functionally overlap with C7. Lesions of these roots are such that compensatory innervation is lost or functionally inadequate, causing the typical clinical dominant C7 entity (8). Because of these exclusive clinical and anatomic characteristics and the small size of the group, which lends itself more easily to analysis, we selected to study those patients with an infrequent dominant C7 lesion.

Imaging

Although there is widespread experience with CT myelography in imaging of cervical roots in OBPLs, a “gold standard” is still lacking. Disadvantages of CT myelography are the need for anesthesia, intrathecal contrast application, and radiation. Several shortcomings of CT myelography are also well known (3, 6, 14, 21, 24). In CT myelography, contrast-filled pseudomeningoceles, subarachnoid space deformity, or missing root shadows are associated with root avulsions but do occur without or may even be lacking in surgically confirmed avulsions. As yet, CT myelography is reported to be conclusive when relevant root shadows can be traced without interruption from the spinal cord to the foramina (6).

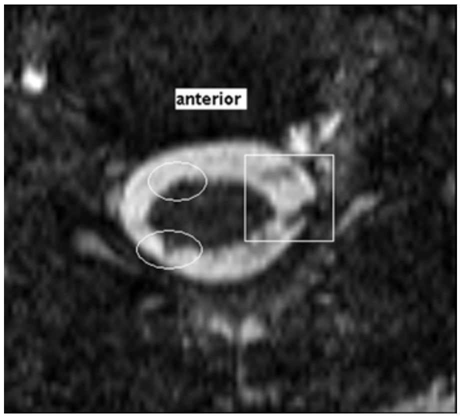

FIGURE 4. *Patient 3. 3D CISS MRI scan, axial view. Clear root shadows on nonaffected side (*square*) and missing root shadows on affected side in absence of a pseudomeningocele. Stumps of anterior and posterior roots with possible recoil (*ovals*).*

3D CISS MRI

MRI studies avoid radiation, are non-invasive, can be performed under mild sedation, are less time-consuming, do not demand anesthesia personnel, and may thus be more cost effective. Nevertheless, introduction of a new technique implies a learning curve for different aspects regarding both technique and interpretation. Despite two minor motion artefacts in our 10 studies, because of slightly too mild sedation, the quality of the studies was good, with images available in the axial and coronal planes. Among eight operative patients, the prediction of root continuity was consistent with the operative findings in six, whereas two remained doubtful. In one patient who recovered and therefore was not operated on, a predicted avulsion would have possibly been incorrect. If in this patient the C7 root had indeed been avulsed, recovery could be explained by reinnervation by adherence to and collateral sprouting from adjacent roots or nearly complete functional overlap of one or more neighbouring roots.

At the cervicothoracic level (roots C8), especially in axial studies, it is sometimes difficult to create such a homogeneous CSF signal that it does not interfere with intradural root images. The quality of coronal imaging is more consistent, probably because in this plane, CSF pulsations do not interfere with the magnetic spin stream, because spins remain within the imaging slab. In this plane, root shadows can be discriminated over their entire intradural length. Precise imaging of the extraforaminal brachial plexus itself in OBPLs requires scanning times of at least half an hour and therefore poses problems in maintaining protocols with only mild sedation (2).

Electrodiagnostics

There are serious doubts about the value of EMG studies in infants with OBPLs (10, 17, 21). A severe clinical picture is often in contradiction to optimistic EMG findings with motor unit potentials suggesting functional innervation (20, 21, 23). In this small selected group, not all data were concordant, for obvious reasons. Furthermore, the EMG studies were performed by several investigators, and of all parameters, only denervation was used. In the dominant C7 lesion, we found persisting signs of denervation, in flaccid or weak triceps muscle or forearm extensors, at approximately 5 months of age in 3 of 10 children studied. This would support the explanation of the appearance in other OBPL groups of so-called inactive motor unit potentials by the concept of polyneural luxury innervation (20, 21, 23). In the dominant C7 lesion, persistent signs of denervation may support the suspicion of a C7 root avulsion or severe nonregenerating neurotmesis, contradicting this concept.

Intraoperative SSEPs, when recorded cervically and cortically, are not diagnostic by themselves but can be useful, when combined with other data, such as preoperative imaging and direct inspection, to form a basis for operative decisions. Although SSEPs may indicate at least partial integrity of dorsal roots, they do not provide any information on the quality and regenerative capacity of the nerve root. Even a clearly avulsed root may show false-positive, reproducible SSEPs. It happens that the proximal stump or the more distal part of an avulsed root will adhere to an adjacent, incompletely damaged root from which sprouting axons may reinnervate the avulsed root. This was observed in two patients who were operated on and may be an explanation for one late spontaneous recovery in which the MRI scan failed to show root shadows of C7. False-positive SSEP recordings may also be caused by EMG artefacts from paraspinal cervical muscles without muscle relaxation or by co-stimulating a contributing branch of the long thoracic nerve at C7 close to its foramen, which forms from branches of multiple spinal nerves at levels close to the foramina. A false-negative response could be caused by inadequate anesthesia (16), technical problems, or at least partially intact but non-conducting intraspinal roots (13).

The functional integrity of an anterior motor root cannot be monitored by SSEP. However, Oberle et al. (13) reported on direct root stimulation close to the foramen and evoked potentials before and after muscle relaxation. Potentials that disappear after muscle relaxation are evoked motor action potentials of spinal muscles. The absence of evoked motor action potentials showed a 100% sensitivity for anterior root lesions as confirmed by intradural inspection in adult patients. This new technique is not yet described in OBPLs.

The observation of obvious motor reactions in three patients after stimulation of clearly avulsed C7 roots is puzzling. At the time of operation, complete Wallerian degeneration should have excluded conduction in motor axons of the C7 myotome. In one patient, the spinal ganglion C7 was encountered very proximal in the foramen and was cut even more proximally. If this was a rupture or elongation of especially the anterior rootlets and not an avulsion, some reinnervation may have been responsible for the observed motor reactions. In another child, the proximal stumps of the motor and sensory branches were found to be adhered to the side of the C8 root, from which collateral sprouting may have caused some regeneration. The third patient showed adherence of the C7 root just distal to the spinal ganglion beside a lesion in continuity with the superior trunk, possibly allowing axonal sprouting toward C7. The stimulation pulse, at the ganglion level, may have been applied too close to the adherence site, bypassing the non-conducting degenerated proximal part.

Cortical motor evoked potentials monitored at spinal roots could add information on the integrity of anterior roots; however, special expertise in infants is required. As is the case in SSEPs, even motor evoked potential-positive recordings would not supply information on the quality, regenerative capacity, and amount of residual rootlets in continuity with their respective root entry zones (19).

Some neurosurgeons prefer directly recorded nerve action potentials by stimulating close to a foramen and record more distally (1, 9). A flat trace is indicative of a complete postganglionic lesion. A preganglionic lesion, in which the sensory nerve still conducts, will show a high amplitude and rapid conduction (50–70 m/s). An intact and regenerating nerve would show small amplitudes and low velocity. We did not perform nerve action potentials because of expected problems in recording signals in the very short (1.5–2.5 cm) supraclavicular plexus structures in infants.

Operation

To expose root avulsions, hemilaminectomy and direct inspection of root entry zones at the spinal cord are still the only way to verify root integrity (3, 14). Any imaging or electrophysiological technique lacking this control therefore cannot be studied conclusively. Even when performed in adult trauma patients, intradural continuous roots, which are thickened or electrophysiologically non-conducting, are encountered, with doubtful regenerative capacity (13). In infants, direct root inspection by (hemi)laminectomy is not performed because of its possible negative effects on the growing cervical spine (15). Next best, therefore, is to rely on intraoperative proof of clearly avulsed spinal nerves with which to correlate imaging techniques and ancillary investigations. Imaging and electrophysiology studies are performed routinely in all our infants with severe OBPLs that meet the criteria for operation. Already in the small Group IV dominant C7 lesion, which is assumed to be most suited to focus investigations on only one root, the shortcomings of advanced diagnostic tools are obvious. In only one of eight patients operated on (Patient 2), the combination of MRI prediction, signs of denervation in the EMG studies, absent motor reaction on direct stimulation of the avulsed C7 root, and absent intraoperative SSEP were consistent with theoretical expectations in the case of a root avulsion.

If, during operation, there is persistent doubt regarding intraspinal continuity or quality of a nerve root, it is advised to consider the root as most probably avulsed. In tailoring the plexus reconstruction, the questionable root can be included in a repair not as a main but merely as a potential source of participating axons.

CONCLUSIONS

Even in the small Group IV dominant C7 lesion, which is assumed to be best suited to focus investigations on only one root, the shortcomings of advanced diagnostic tools remain obvious. In the other, larger OBPL groups with multilevel root pathological findings and more functional overlap, these shortcomings become even more accentuated. With the new 3D CISS MRI technique described, excellent imaging quality helps to delineate the lesions and tends to be a reliable tool to predict avulsions, although the precise diagnosis of cervical root continuity or avulsion in OBPLs is still not possible as long as there is no direct operative intradural visual control of roots and root entry sites at the spinal cord. The combination with conventional electrophysiological investigations has restricted value in the clinical decision making in obstetric brachial plexus surgery. However, recently reported new electrophysiological techniques might be more promising.

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CHAPTER 5

SECONDARY DEFORMITIES OF THE SHOULDER IN INFANTS WITH AN OBSTETRIC BRACHIAL PLEXUS LESION CONSIDERED FOR NEUROSURGICAL TREATMENT

Based on:

Secondary deformities of the shoulder in infants with an obstetric brachial plexus lesion considered for neurosurgical treatment

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Secondary deformities of the shoulder in infants with an obstetrical brachial plexus lesions considered for neurosurgical treatment

OBJECT

The authors performed a prospective study in which magnetic resonance (MR) imaging was conducted in 26 consecutive infants (mean age 5.6 months, range 2.7–14.5 months) in whom recovery from an obstetric lesion of the brachial plexus had been inadequate in the first 3 months of life. The purpose was to identify early secondary deformations of the shoulder in obstetrical brachial plexus lesions (OBPLs).

METHODS

Features of the shoulders were analyzed according to a standardized MR imaging protocol in patients with OBPLs. Measurements were made of the appearance of the glenoid, glenoid version, and the position of the humeral head.

The appearance of the glenoid on the affected side was normal in only 11 shoulders. In the remainder it was convex in eight and biconcave in seven cases. The degree of humeral head subluxation was significantly greater ($p = 0.001$) in affected shoulders than in normal shoulders (152 and 170°, respectively). The presence of abnormal glenoid retroversion and humeral head subluxation increased with age: there was a statistical difference ($p = 0.001$) between infants younger than 5 months of age and those who were older.

CONCLUSIONS

Magnetic resonance imaging demonstrates shoulder-related anatomical and nerve root lesion, allowing evaluation of neural, osseous, and cartilaginous structures in younger children.

In most children with an OBPL, improvement of the neurological deficits can be expected within the 1st year of life.[1,9,15,18] At present neurosurgical treatment is considered in infants with (sub)total lesions and those without biceps muscle function at 3 months of age.2,6 In these infants neuroimaging is part of the preoperative evaluation and is usually performed at 4 months of age. This examination focuses on diagnosing the type of lesions — that is, distinguishing neurotmesis from avulsion because regenerating axons cannot be expected in preganglionic spinal nerve root avulsions.[1]

Abbreviations used in this chapter
MR = magnetic resonance;
OBPL = obstetrical brachial plexus lesion.

For the diagnosis of brachial plexus lesions in infants, MR imaging has surpassed computerized tomography myelography as the modality of choice. In applying different techniques in a non-invasive way, high-power MR images reveal plexus structures with great detail.[5,10,17]

Although clinical symptoms are primarily caused by the brachial plexus lesion in infants with an OBPL, secondary deformities of the upper extremity may develop and influence the symptoms, predominantly around the shoulder. Information regarding these secondary shoulder deformities can be acquired during the MR imaging procedure by targeting the relevant neural structures.

Because brachial plexus lesions predominantly affect the upper cervical nerve roots or trunk

(C5–6), muscular imbalance results around the shoulder, with paralysis of the abductors and external rotators and relative dominance of internal rotators.[1] Consequently, a characteristic secondary shoulder deformity develops consisting of a flexion and internal rotation contracture of the shoulder as well as a posterior subluxation of the humeral head[1-3] (referred to as “humeral head subluxation”).

It has been shown that secondary structural shoulder deformities develop early in life and may persist despite improvement in neurological status.[16] Although these secondary deformities may be compensatory lesions resulting from the altered muscular balance around the shoulder, in some cases these persistent deformities may even worsen the functional impairment caused by a residual neurological deficit.[1]

The prevalence of shoulder deformities has been reported in 40 to 70% of cases.[1,7,8,13,19] Until recently standard radiography and computerized tomography scanning were conducted to analyze the developing shoulder deformities. Their shortcoming is that they focus on the osseous structures. In this respect MR imaging better visualizes the predominantly cartilaginous shoulder in young children.

To assess the prevalence of secondary deformities of the shoulder in infants with OBPLs considered for neurosurgical reconstruction, we performed a prospective MR imaging–based study.

CLINICAL MATERIAL AND METHODS

Patient Population

This prospective study was based on a group of infants with OBPLs who were being considered for reconstructive procedures because of inadequate neurological recovery in the first 3 months of life. Twenty-six consecutive children with a unilateral OBPL (16 boys and 10 girls; mean age 5.6 months [range 2.7 – 14.5 months]; 11 left and 15 right-sided lesions) were evaluated between July 1998 and September 2000. In all cases the cervical spine and shoulders bilaterally were examined using MR imaging.

The severity of the neurological deficit was classified according to the system of Narakas[11] (Table 1).

Magnetic Resonance Imaging Protocol

For the MR imaging investigation (Siemens Magnetom 1.5-tesla Vision; Siemens, Erlangen, Germany), the children received an intramuscular cocktail of pethidine, droperidol, and chlorpromazine. They underwent electrocardiography, oxygen saturation, and video monitoring. To visualize both shoulders a three-dimensional fast imaging with steady-state precession pulse-acquisition sequence imager (TR 25 msec, TE 10 msec, flip angle 40°) with 1.5-mm partitions was used. Measurements of both shoulders were performed in the axial plane at the midglenoid level.

Three established measurement methods were used in this analysis. In the first, the glenoid form was classified qualitatively as concave-flat, convex, or biconcave according the system proposed by Birch, et al.,[1] (Fig. 1).

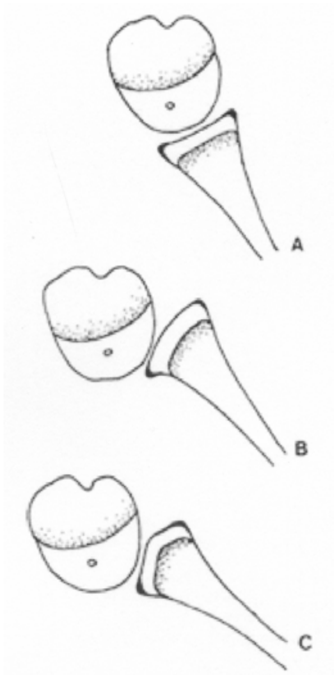

Fig. 1. Diagrams showing qualitative classification of concave flat (**A**), convex (**B**), and biconcave (C) glenoids. The British editorial society of Bone and Joint Surgery. Reprinted with permission from **J Bone Joint Surg Br 83:**551–555, 2001.

TABLE 1

Summary of Narakas classification of OBPLs

Narakas Group	Neurological Signs
1: C5–6 lesion	paresis of deltoid & biceps muscles
2: C5–7 lesion	paresis of shoulder & extension of elbow, wrist & fingers
3: C5–8 lesion	paresis virtually complete w/ some finger flexion
4: whole plexus involvement	paresis of arm; Horner sign

TABLE 2
*Summary of neuroimaging findings**

Case No.	Age (mos) at MRI, Sex	Narakas Group	Affected Side	Glenoid Form† (affected side)	Glenoid Version ‡ Affected Side	Glenoid Version ‡ Normal Side	Humeral Head Sublux (°)§ Affected Side	Humeral Head Sublux (°)§ Normal Side
1	2.7, F	2	rt	1	−7	−3	175	169
2	3.2, F	1	lt	1	−10	−9	167	160
3	3.8, M	2	rt	1	−6	−9	177	172
4	3.9, M	3	rt	2	−10	0	154	156
5	4.1, M	2	lt	1	−13	−8	166	159
6	4.1, M	2	lt	3	−21	−5	124	170
7	4.4, F	1	rt	1	−8	−17	180	193
8	4.5, M	2	rt	1	0	1	176	174
9	4.6, F	2	lt	3	−18	2	131	175
10	4.6, M	4	rt	2	−5	−6	160	178
11	4.7, M	4	rt	3	−4	−7	138	176
12	4.8, F	1	lt	1	−6	−23	171	151
13	5.0, M	1	lt	1	−9	−1	174	161
14	5.0, M	1	lt	1	−17	−9	146	156
15	5.0, M	1	rt	2	−30	−4	102	167
16	5.0, M	1	lt	3	20	15	142	173
17	5.1, F	1	lt	1	2	9	178	200
18	17.5, M	1	rt	3	−25	−11	126	179
19	5.4, M	2	lt	2	−10	−2	159	162
20	5.7, F	1	rt	2	−19	0	151	164
21	5.7, F	2	rt	2	0	−2	157	180
22	5.9, F	3	rt	2	−27	−15	128	170
23	6.9, M	1	rt	2	−17	−17	146	168
24	8.7, F	1	rt	3	−24	−10	134	156
25	13.8, M	2	rt	3	−18	−3	135	179
26	14.5, M	2	lt	1	−15	−6	152	160

* Sublux = subluxation.
† Glenoid form: 1, concave-flat; 2, convex; 3, biconcave.
‡ Glenoid version according to Friedman, et al.
§ Humeral head subluxation according to Papilion and Shall.

In the second method, the glenoid version angle was measured according to that described by Friedman, et al.,[4] — that is, the angle between the line connecting the base of the anterior and posterior labrum and the line connecting the middle of the glenoid and the medial margin of the scapula (Fig. 2). By definition 90° was subtracted from the aforementioned angle to determine the glenoid version angle. A negative value indicated a retroverted glenoid. Finally, in the third method, the subluxation of the humeral head was measured using a modification of the method described by Papilion and Shall;[12] that is, the angle between the line medial margin of scapula/midpoint glenoid and the line midpoint glenoid/center of humeral head.

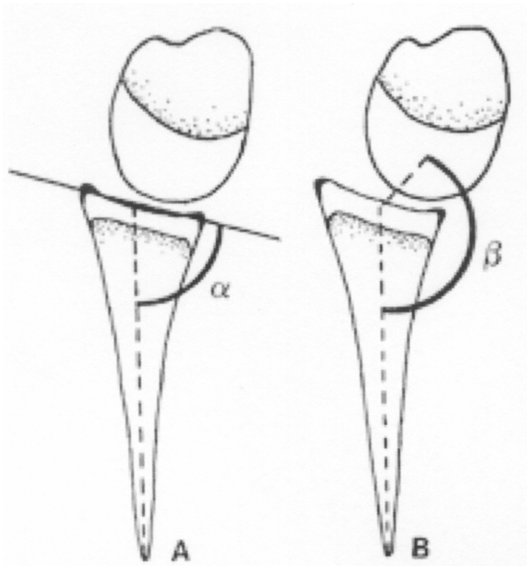

Fig. 2. Diagrams. **A:** The method of measuring the glenoid version angle according to Friedman et al. The angle in the posterior quadrant is measured, and 90° is subtracted from this angle to determine glenoid version. B: The method of measuring the posterior subluxation of the humeral head according to Papilion and Shall The British editorial society of Bone and Joint Surgery. Reprinted with permission from **J Bone Joint Surg Br 83:**551–555, 2001.

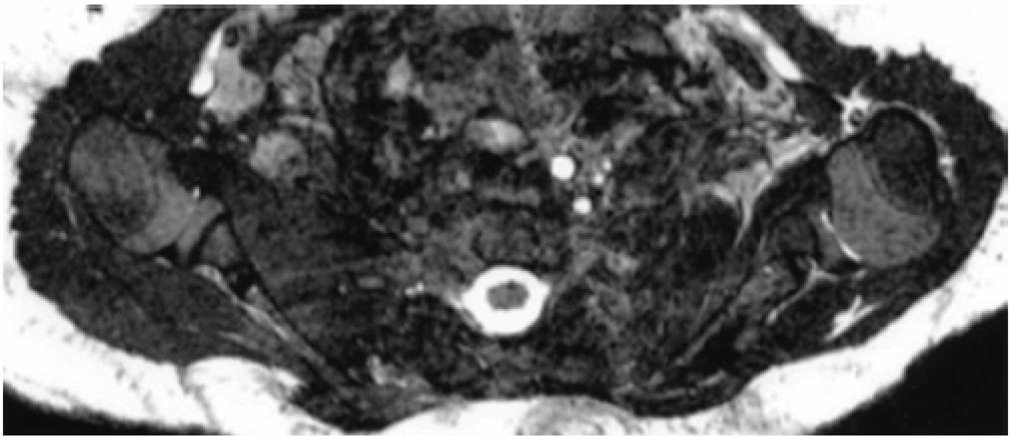

Fig. 3. Case 16. Axial fast imaging with steady-state precession scan. On the right side there is a convex glenoid and subluxation of the humeral head. The contralateral shoulder is normal.

Statistical Analysis

All measurements were made on images in digital format by using post processing software (Radworks 4.0; Applicare Medical Imaging BV, Zeist, The Netherlands) Statistical significance of the differences between the pathological and normal side was tested using parametric tests for quantitative data and the Fisher exact test for nominal data.

RESULTS

There were no complications related to the sedation. All MR imaging studies were of adequate quality. Detailed patient-related data are presented in Table 2.

The glenoid form on the affected side was qualitatively normal in 11 shoulders. Pathological glenoids were convex in eight cases and biconcave in seven (Fig. 3). In addition the extent of humeral head subluxation was significantly greater (p = 0.001) in the shoulders on the affected side (152° compared with 170°, respectively) and the glenoid version was different from the contralateral side (-11.4° and -5.4°, p = 0.004). There was no relation between shoulder deformities and Narakas grouping. Post-traumatic abnormalities were not seen.

Glenoid retroversion and humeral head subluxation increased as the patient aged. The influence of age became apparent when comparing younger (< 5 months) and older infants (< 5 months) within the study group. In infants younger than 5 months, the glenoid version angle and extent of humeral head subluxation of affected and normal shoulders were not significantly different (Table 3). In infants of 5 months of age and more, however, the extent of glenoid version and humeral head subluxation were significantly different compared with the unaffected side (p = 0.001; Table 3).

TABLE 3

*Quantitative measurements of shoulders**

	Patient Group					
	<5 Mos Old			≥5 Mos Old		
	Affected Side	Healthy Side	p Value	Affected Side	Healthy Side	p Value
glenoid version	−9°	−7°	NS	−3.5°	−4°	0.001
amount of sublux	160°	169°	NS	145°	169°	0.001

* NS = not significant.

DISCUSSION

Modern neurosurgical techniques enable early intervention in cases of OBPL in infants with incomplete recovery within the first 3 to 6 months of life and infants with a subtotal lesion. Magnetic resonance imaging has been introduced to visualize the plexus lesion. This modality is also suitable for visualizing the early secondary shoulder deformities that may be present in some infants.16 This is confirmed in the present study, revealing the secondary deformities of cartilaginous structures of the shoulder in most within the first 14 months of life. We found in the majority of infants 5 months of age and older that the glenoid form is abnormal, glenoid version is altered, and humeral head subluxation is present. In our study, a substantial number of patients were selected to undergo imaging because of a persistent neurological deficit; however, we think that secondary shoulder deformities are not limited to patients with such a severe type of OBPL. This rationale is supported by Fairbank,[3] who reported shoulder deformities in 28 children in a consecutive group of 37 OBPLs involving all types of brachial plexus lesion. In addition, the authors of two recent studies reported a persistent internal rotation contracture of the shoulder in most infants with OBPL in whom there was a delay of more than 2 months in biceps muscle recovery.[9,18]

We found no evidence to support the hypothesis of some authors that secondary shoulder deformities in OBPL are caused by a fracture of the proximal humeral epiphysis, which leads to an increased humeral head retroversion.[14,20] The clinical relevance of the shoulder deformities is still unclear. Are these deformities adaptations of skeleton to the altered muscular function due to the complete or incomplete OBPL and, as such, compensatory mechanisms? Alternately, are the deformities dysfunctional and responsible or partly responsible for loss of function in some children? Perhaps there is no clear answer but deformities may differ in their role in different children. In our view these deformities alone are seldom entirely responsible for clinical symptoms.

CONCLUSIONS

Our findings indicate that because MR imaging reveals both damage to the nerve roots and the anatomy of the shoulder, MR imaging allows evaluation of the neural, osseous, and cartilaginous structures in the younger child. Accordingly, MR imaging studies of both shoulder and cervical spine should routinely be performed in one session in patients being considered for neurosurgical intervention. Early detection of shoulder deformities is possible and thereby opens possibilities for new types of treatment.

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CHAPTER 6

CENTRAL ISSUE IN ACCESSORY NERVE TO SUPRASCAPULAR NERVE TRANSFER TO RESTORE SHOULDER EXOROTATION IN OTHERWISE SPONTANEOUSLY RECOVERED OBSTETRIC BRACHIAL PLEXUS LESIONS ?

Accepted for publication in Neurosurgery

CENTRAL ISSUE IN ACCESSORY NERVE TO SUPRASCAPULAR NERVE TRANSFER TO RESTORE SHOULDER EXOROTATION IN OTHERWISE SPONTANEOUSLY RECOVERED OBSTETRIC BRACHIAL PLEXUS LESIONS ?

SUMMARY

A group of 54 children with an obstetric brachial plexus lesion (OBPL) showed satisfactory spontaneous recovery of shoulder and arm function except for active shoulder exorotation. Functional exorotation was achieved in 50 children by performing an accessory to suprascapular nerve transfer. Because all other C5 and C6 or superior trunk functions spontaneously recovered, spinatus muscles did not show extensive wasting on preoperative MR-studies or denervation signs on electromyography, the suprascapular nerves proved reactive at intra-operative electro-stimulation and histological investigation was normal, the authors conclude that although the described nerve transfer is effective the cause for the lack of recovery of exorotation is rather a form of developmental apraxia caused by defective motor programming at a critical age than a residual or isolated peripheral nerve problem. The importance of the article is to point to central nervous system changes or aberrant adjustment after OBPL as possible main cause for lack of spontaneous recovery of certain functions.

INTRODUCTION

An obstetric brachial plexus lesion (OBPL) is a stretch injury to the brachial plexus and can be classified according to Narakas (table 1) (15). Microsurgical reconstruction is indicated in severe lesions with insufficient spontaneous recovery. In Group I lesions, with damage of C5 and C6 or Superior Trunk (table 1), shoulder function, elbow flexion and forearm supination are more or less limited. The main criterium for surgery is a failure to recover biceps function at four months (4, 7, 17, 22). In a number of children spontaneous recovery of biceps function at four to six months may be sufficient as not to fulfil the criteria for surgery. However, a striking and complete lack of active shoulder exorotation persists despite ongoing improvement of other shoulder and arm functions. It is poorly understood why suprascapular nerve (SSN) and infraspinatus muscle function, as expressed by shoulder exorotation do not recover, while other functions related to the superior trunk do. To restore active shoulder exorotation, an accessory nerve (XIN) to SSN transfer is a treatment option (3, 8, 13, 17, 20, 26). However, if performed as part of the primary reconstruction for an OBPL results tend to be disappointing (17). The SSN is a pure motor nerve that innervates the infra- and supraspinatus muscles of which the first is the main shoulder exorotator. It is a branch of the superior trunk, which forms from both anterior and posterior divisions (29) of the fifth and sixth cervical nerve roots. We report on the results and follow-up of 54 children, who underwent a XIN to SSN transfer as a primary and single procedure or as a separate secondary procedure. Possible causes of failure to spontaneously recover active shoulder exorotation are discussed.

MATERIALS AND METHODS

Fifty-four children with a Group I OBPL treated between 1998 and 2004 were included in our study. Forty-five children were presented and treated at the VU University Medical Centre in Amsterdam, the Netherlands and nine at the Children's Hospital in Linz, Austria. Of this group 47 children had sufficient recovery of biceps and deltoid function at four to six months to justify conservative treatment. During a baseline phase prior to operative treatment all children received an intensive program of weekly physiotherapy and daily exercises directed at stimulation of active shoulder exorotation and preserving passive shoulder mobility. However, at repeated measurements, where subjects served as their own controls, they failed to recover any active exorotation. From the age of three or four months, the children were followed at intervals of four months till ages of 10 months and older before operative treatment by XIN to SSN

Classification OBPL	Lesion	Clinical presentation
Group 1	C5, C6 or Superior Trunk	Paralysis of deltoid, shoulder exorotation and biceps
Group 2	C5, C6 or Superior Trunk, C7 or Medial Trunk	Idem Group 1, with paralysis of extension of elbow, wrist and digits. Typical 'waiter tip' hand
Group 3	C5, C6 or Superior Trunk, C7 or Medial trunk, Incomplete C8, T1 or Inferior Trunk	Except for some finger flexion, virtually complete paralysis of upper extremity
Group 4	Complete plexus	Complete flaccid paralysis of upper extremity Horner Sign
Group 4 –dominant C-7	C7 or Medial Trunk, Incomplete other plexus Structures	Paralysis of shoulder adduction and elbow extension and mostly in different degree of wrist and finger extension. Varying incomplete other muscle deficits.

Table 1. Narakas classification of obstetric brachial plexus lesions

transfer was considered and performed. Seven of the 54 children, fulfilling the criteria for early surgery underwent brachial plexus exploration between the fourth and sixth months of age (4, 7, 22). In one a neurolysis and in six reconstruction with grafts from C5- and/or C6-nerve roots to the distal superior trunk and SSN were performed after resection of large traumatic neuromas. Because recovery of shoulder and arm function was comparable to the spontaneous recovery in the 47 conservatively treated patients but without improvement of exorotation, these operative cases were included. These seven children were followed for at least a year after initial surgery before deciding to perform a XIN to SSN transfer as a separate, secondary procedure. All patients were followed and operated upon by the same pediatric neurosurgeon (WJRvO). Postoperative follow-up was every four months during the first year, every six months during the second year and at the end of the third year.

The following patient characteristics were registered: gender, side of the lesion, first or later delivery, type of delivery, position of the child during delivery, complications during pregnancy, associated birth lesions, birth weight and age at first presentation in our brachial plexus clinics. The results of 3D-CISS (continuous interference in steady state) MR-scans (16) and of needle electromyography performed in children with still insufficient recovery in the fourth month, previous plexus exploration, patient's age at the time of surgery, whether or not a combined shoulder release had been performed and the response of infra- and supraspinatus muscle after intra-operative electro-stimulation of the SSN were also noted. In three patients MR-scans of the shoulder were performed to compare muscle trophy of relevant infra- and supraspinatus muscles to the non-affected side. In two of those, MR-scans were made 4 and 3 years before the nerve transfer and in all three children one day before and two years after the nerve transfer.

Two investigators (neurosurgeon and physiatrist) scored degrees of passive and active shoulder exorotation (normal range – 70 to 80 degrees) with the upper arm gently held in adduction by the investigators and the elbow actively flexed in 90 degrees, degrees of active abduction (range 0 to 160 degrees), forearm supination (range 0 – 80 degrees) and passive elbow extension. Active motion was always scored against gravity, with the child in the sitting or standing position. First clinical evidence of active exorotation was documented, as well as the shortest postoperative interval to reach the maximal individual result for shoulder exorotation and abduction.

Shoulder function was additionally scored using the modified Mallet scale for abduction, exorotation and hand-to-mouth movement (figure 1) (7) (In infants the hand-to-head and hand-to-back movements are difficult to provoke and were therefore not assessed).

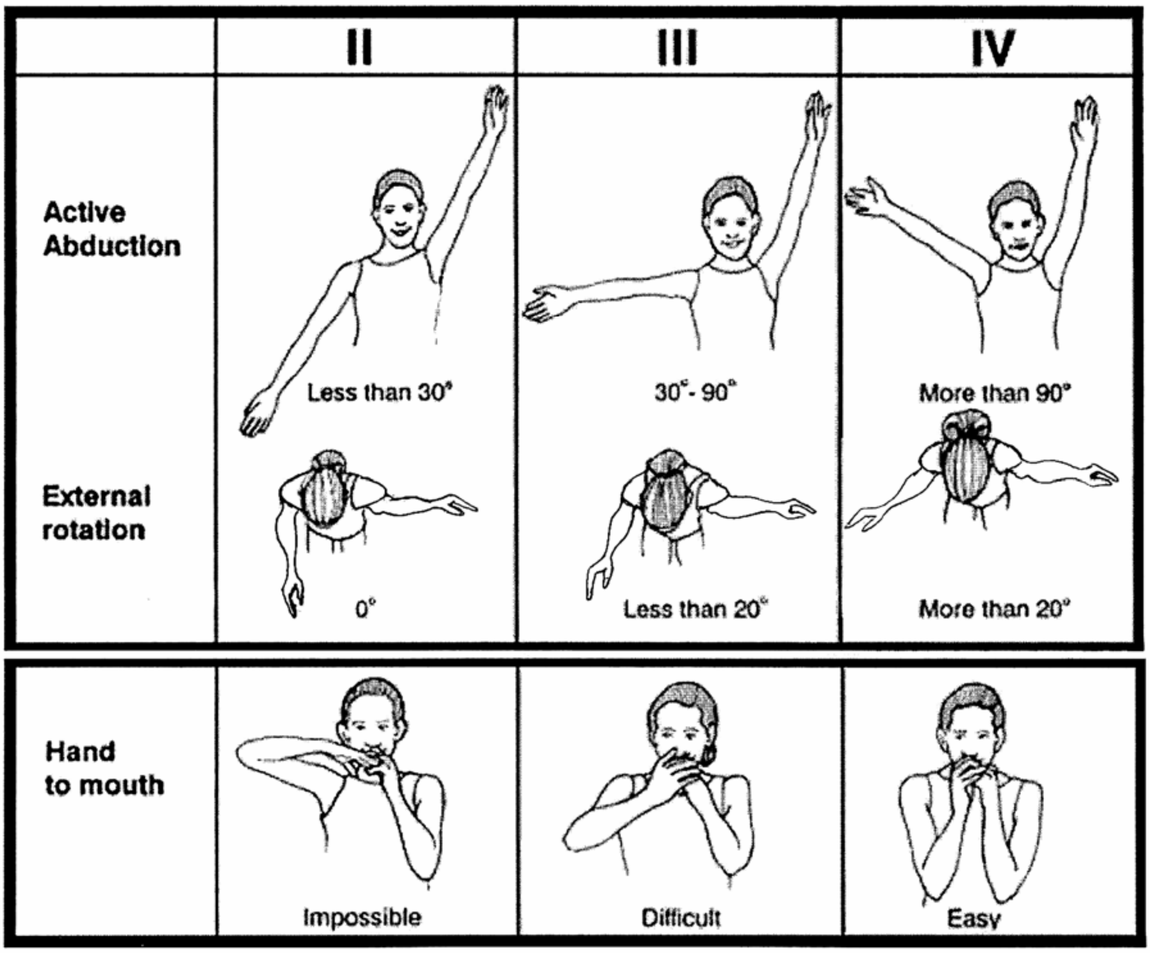

Figure 1
Modified Mallet scale for shoulder function. The ‘Trumpet-sign’ is demonstrated in the hand-to-mouth scale with score II and less pronounced III. The hand-to-head and hand-to-back scales are left out.

Cumulative survival curves as a function of time to reach a certain threshold in active exorotation were generated with the Kaplan-Meier method. Several thresholds (0, 20 and 40 degrees) were analysed. Patients who did not reach that threshold during follow-up were censored at the time of the last visit. Differences between subgroups with respect to reaching these thresholds were compared using the log-rank test. Statistical significance was defined by $p< 0.05$. All statistical analysis were performed with SPSS for Windows.

Operative procedure

Through a three centimetre, supraclavicular skin incision the XIN was explored from the upper anterior rim of the trapezius muscle, then traced and a branch cut distally from the muscle, transferred and neurotized with fibrin glue to the SSN, which was cut just after branching off from the superior trunk. Electrical nerve stimulation (single use Neuro-PulseTM surgical nerve locator, Bovie Aaron Medical, St Petersburg, USA) with stimulus at 0.5 mA or 1 mA was used to check for motor respons in the infra- and supraspinatus muscles and the trapezius muscle before severing the nerves. Reaction was positive when movement of the shoulder occurred and muscle contraction was palpated over the spinatus muscles. To document the procedure, additional intra-operative needle electro-myographic responses in infra- and supra-spinatus muscles were recorded and histology was performed of the proximal part (0.5 cm) of the suprascapular nerve in five patients. One patient previously underwent grafting from C6 to the distal superior trunk and suprascapular nerve in the presence of an avulsion of the fifth cervical nerve, the other four patients were thus far treated conservatively.

If there was a more than 50 degrees restriction of passive motion for exorotation with the upper arm in adduction, a shoulder release by subscapular tendon lengthening and reposition

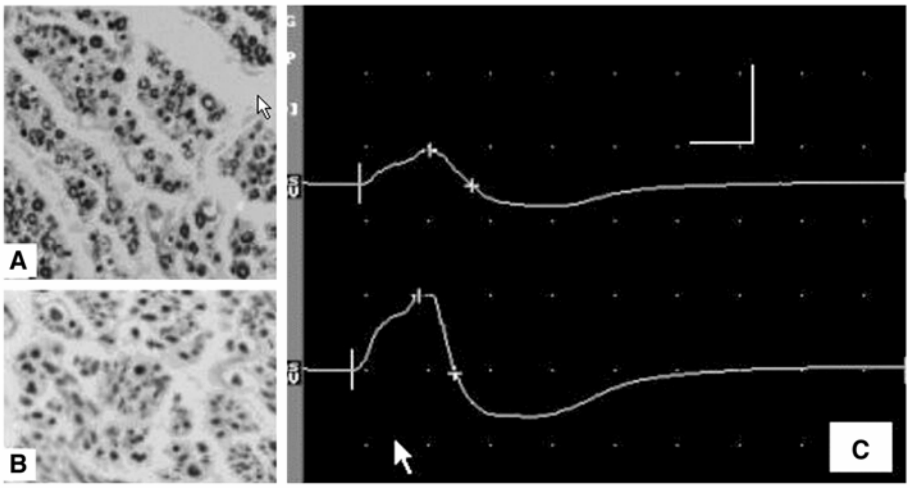

Figure 2
Specimen of proximal suprascapular nerve of two patients obtained during accessory to suprascapular nerve transfer.
A) Klüver stain, showing normally sized myelinated nerve fibres in a child aged 16 months, previously conservatively treated.
B) Bodian stain, showing normally sized axons in a child aged 6.5 years, 6 years after grafting the sixth cervical root to the superior trunk
C) Intra-operative needle electromyography in the same patient. Motor response in supra- (upper) and infraspinatus muscle after single stimulation of the suprascapular nerve (calibration: 3 msec and 5 mV per division).

of the humeral head was performed simultaneously (25). The head, shoulder and arm were immobilised for three weeks in a scotchcast plaster with the arm positioned in about 30 degrees abduction and different degrees of exorotation, depending on the possible passive mobility and to prevent worsening of the range of passive endorotation (23, 24, 25). Procedures take around 45 minutes and patients can be released on the day of operation. Instructions were given for weekly physiotherapy and daily exercises by the parents to prevent endorotation contractures and to stimulate active exorotation.

RESULTS

Patient characteristics are summarized in table 2. Most (40/54) children affected were born from multiparous mothers. Pregnancy was complicated by diabetes in two and hypertension in one, whereas one mother underwent appendectomy in the twentieth week. Duration of pregnancy ranged from 32 to 43 weeks (mean: 39.5 and median: 40 weeks). Birth weight ranged from 1670 grams to 5080 grams, (mean: 4058 grams, median: 4165 grams). Associated lesions consisted of isolated clavicula fractures, (five homolateral, one contralateral, one bilateral) or in combination with a hematoma in the sternocleido-mastoid muscle (one), a phrenic nerve lesion (one) or bilateral OBPL (one). Other isolated lesions were hematomas in the sternocleidomastoid muscle (four) or the affected arm (one). The age at first presentation of children in our plexus clinics ranged from 1 to 75 months (mean: 7.0 months, median: 3.0 months). Electromyography at four months, performed in 40 patients, showed signs of denervation of spinatus muscles in one patient. The 3D-CISS-MRI studies showed pseudomeningoceles at relevant levels of C5 and/or C6 in 21 of 40 patients. The age at which an accessory to suprascapular nerve transfer was performed ranged from 6 to 84 months (mean: 21.7 months, median: 15.0 months). To relieve an internal rotation contracture a simultaneous, combined shoulder release was necessary in 14 patients (mean age 28.9 months, median 19.5 months, range 9 to 80 months). At electro-stimulation of the suprascapular nerve, the majority of patients (81% or 44 patients) showed a clear motor reaction in the infra- and supraspinatus muscles, in six patients the response was not documented, although electro-stimulation is routinely performed. Four patients showed no response. Intra-operative electromyography of the infra- and supraspinatus muscles, performed in five cases, showed normal muscle action potentials (figure 2). Histology of the suprascapular nerve in the child who underwent plexus reconstruction and grafting earlier, showed normal myelination. The same holds true for four children that were thus far treated conservatively (figure 2).

GENDER		LESION SIDE		CHILD NUMBER		BIRTH WEIGHT MEAN 4051 g MEDIAN 4160 g		POSITION		DELIVERY		PREGNANCY DURATION MEAN 39,5 weeks MEDIAN 40,0 weeks	
female	male	left	right	first	≥ 2nd	≤ 4 kg	> 4 kg	head first	breech	spontaneous	instrumental	≤ 40 w	> 40 w
25	29	19	35	14	40	22	32	44	10	39	15	37	17

PREGNANCY COMPLICATIONS		ASSOCIATED BIRTHLESION(S)		3D-CISS-MRI				AGE AT OPERATION N.XI – N.SS MEAN 21.7 months MEDIAN 15.0 months		SHOULDER RELEASE COMBINED		REACTION MUSCLE TO ELECTROSTIMULUS		
Yes	no	yes	no	normal	lesion C5 or C6	lesion not C5 or C6	not performed	≤ 12 months	> 12 months	yes	no	yes	no	?
4	50	15	39	21	10	8	15	15	39	14	40	44	4	6

Table 2. Characteristics of 54 children with obstetric brachial plexus lesions and residual lack of active shoulder exorotation.

ACTIVE EXOROTATION		-70^0 - -50^0	$\geq -50^0$ - -20^0	$\geq -20^0$ - 0^0	$\geq 0^0$	IMPROVED
Number of patients at operation	54	54	0	0	0	-
Number of patients at 4 months FU	54	15	5	6	28	39/54
Number of patients at 8 months FU	52	4	1	4	43	48/52
Number of patients at 12 months FU	47	1	1	1	44	46/47
Number of patients at 24 months FU	30	0	0	1	29	30/30

Table 3a. Follow up (FU) in degrees (x^0) of exorotation with arm in adduction in patients after accessory to suprascapular nerve transfer. At operation exorotation was -70^0 in all patients.

ACTIVE EXOROTATION	TIME IN MONTHS
0^0	4
20	12
30	18
40	24

Table 3b. Median time in months to reach functional exorotation

	$< 0^0$ EXO	$\geq 0^0$ - $< 20^0$ EXO	$\geq 20^0$ - $< 40^0$ EXO	$\geq 40^0$ EXO	MALLET EXO II	MALLET EXO III	MALLET EXO IV
Number of patients : n=54	4	11	15	24	4	11	39
Mean first max. exo in months	12	10	14.7	17.2	-	16.6	12.2
Median first max. exo in months	10	18	12	18	-	8	12
Range to reach first max. exo in months	4-24	8-36	8-24	8-36	-	4-12	4-36

Table 4. Results at last follow-up visits of active exorotation (exo) in 54 children after accessory to suprascapular nerve transfer.
Note: as postoperative follow up ranged from 4 months to several years, individual maximum (max) values for exoration are not fixed, and may change during further follow up (See figure 3: survival curve).

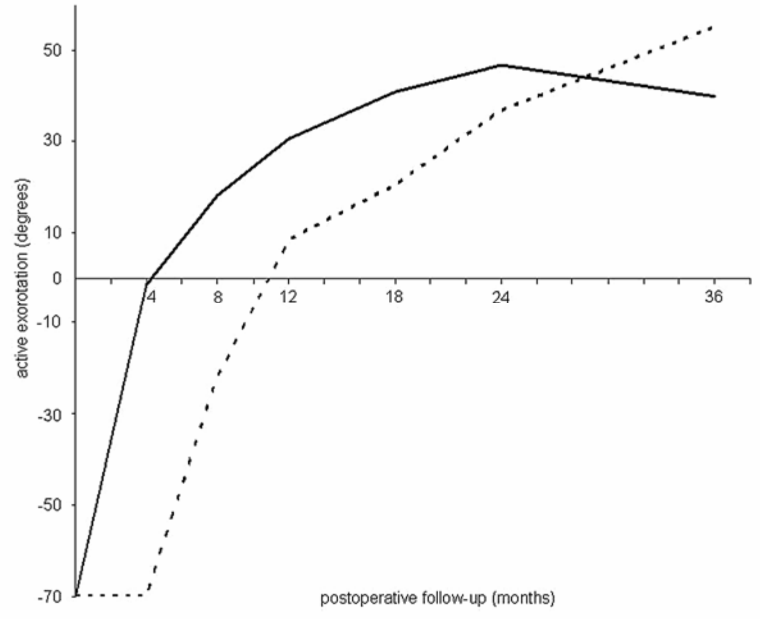

Figure 3
Active exorotation during follow-up after accessory to suprascapular nerve transfer. One group (39 patients) shows early recovery at 4 months (straight line), another (15 patients) showed delayed recovery (dotted line). The end results of both groups are comparable.

Duration of follow-up was 36 months in 15 children, 24 months in 15 children, one to two years in 17 children and less than one year in 7 children.

Recovery of active exorotation

Effects of the nerve transfer on active exorotation during postoperative follow-up are shown in tables 3 and 4 and figures 3 and 4. At four months follow-up 39 patients (72%) showed at least 20 degrees improvement of active exorotation.

At eight months follow-up four patients of 52 (8%) and at 12 months one patient of 47 (2%) did not yet show any improvement. All four patients who did not show signs of recovery at 8 months had a total follow-up of at least 24 months. In 39 patients recovery was already present at the first follow-up visit, four months after surgery. In 15 patients recovery was delayed, however without affecting the ultimate recovery.

Table 4 shows the categories of maximally reached shoulder exorotation at respective mean and median times of postoperative follow-up. The time to reach functional results for exorotation (range of motion improving from -70 to at least 0 degrees or more) for the median of the study group are represented in table 3b. Of the total group of 54 children, four patients had insufficient active exorotation, that is less than 0 degrees at their final follow-up visit. However, of these, one child improved after secondary release of a passive shoulder deficit.

None of the characteristics listed in table 2 significantly affected the time to reach any of the analysed thresholds in active exorotation except for instrumental delivery to reach 0 or 20 degrees ($p<0.05$, log-rank test) There was no significant difference in response to the nerve transfer for different age groups. Children younger than 12 months (15 patients) or over 12 months (39 patients), or younger than 18 months (33 patients) or over 18 months (21 patients) of age at operation reached similar active range of exorotation within corresponding time of follow up. The six children who underwent the nerve transfer as secondary procedure after primary plexus reconstruction showed a tendency not to reach end values over 40 degrees exorotation. Two patients reached 20-40 degrees, two other 0-20 but degrees and two, who had a short follow up of less than / or 8 months, still did not reach 0 degrees. The child that underwent a neurolysis as primary procedure, reached 70 degrees of active exorotation.

Passive shoulder exorotation

At the end of follow up 12 patients had a passive range of motion of 45 degrees or less

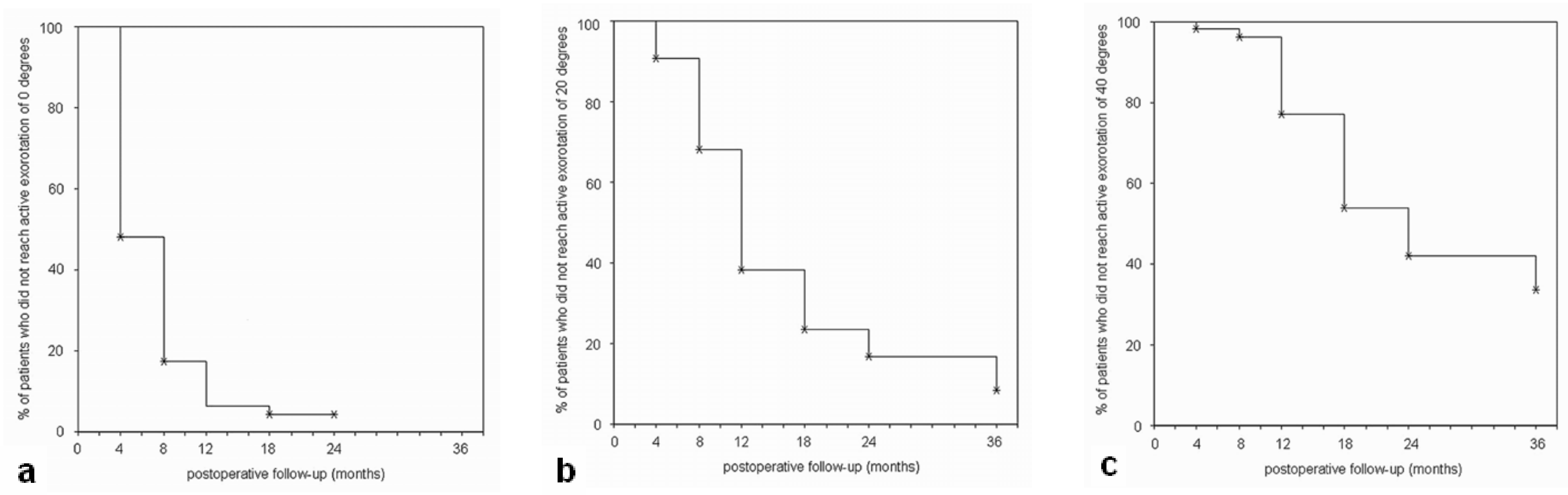

Figure 4 Survival curves showing percentage of patients not reaching
a) 0 degrees
b) 20 degrees
c) 40 degrees
of active exorotation during follow-up after accessory to suprascapular nerve transfer.

ABDUCTION AT TIME OF OPERATION	(n)	**ABDUCTION AT LAST FOLLOW UP**				
		≤ 40^0	**> 40^0 - < 90^0**	**90^0**	**> 90^0 - ≤ 120^0**	**> 120^0**
≤ 40^0	2	1	1			
>40^0 - <90^0	6	1	3	1		1
90^0	14			5	6	3
>90^0 - ≤ 120^0	16				4	12
> 120^0	16					16*
Total	54	2	4	6	10	32

Table 5. Active abduction against gravity in degrees at operation and at last follow up after accessory to suprascapular nerve transfer. In total 27 patients improved of which * 3 patients improved 30 degrees or more within the best group (>120^0)

(range 10 – 45 degrees). In one child a serious passive deficit of 70 degrees (range -70 - 10 degrees) seemed to restrict obtained active motion to -20 degrees. After a shoulder release performed at 24 months follow-up, the child showed improvement of active exorotation to 0 degrees and passive exorotation to 60 degrees.

Active shoulder abduction (table 5)

At the time of operation 46 (85%) children had active abduction of at least 90 degrees. During follow-up abduction improved in 27 children (50%) and remained unchanged in 26 (48%). Active abduction of less than 90 degrees persisted in six patients. In one child abduction worsened from 45 to 30 degrees. *Active supination of the forearm* showed a deficiency of 20 to 45 degrees in nine patients, and over 45 degrees in two patients. Most children (80% or 43), had a near to, or normal range of supination from 0 to 60 or 80 degrees.

Passive extension deficits of the elbow were present in 15 patients and ranged between 5 and 10 degrees in nine patients, between 10 and 20 degrees in four and was over 20 degrees in two of the 54 children. Except for two children, there were no combined severe deficiencies for range of movement for active exorotation, active supination or passive elbow movement. One child had a combined deficiency of 30 degrees for passive elbow extension and active supination (range 0 to 50 degrees) but had an improved active exorotation to 45 degrees.
Another child had this combination with 20 degrees deficiency for elbow extension, 40 degrees for supination (range 0 to 40 degrees) but also a bad result with only -20 degrees for active exorotation (the same child as mentioned under passive exorotation).

Mallet score In table 6 the Mallet score registered just before operation and at individual end of follow-up is represented. In the hand-to-mouth movement a more or less pronounced Trumpet-sign is still present in 17 patients. The scores for abduction and exorotation are congruent to the values in tables 4 and 5.

MALLET SCORE	**AT TIME OF OPERATION**			**MAXIMUM RESULT**		
	II	**III**	**IV**	**II**	**III**	**IV**
Abduction	1	21	32	0	12	42
Exorotation	54	0	0	4	10	40
Hand to mouth	54	0	0	7	10	37

Table 6. Mallet scores for shoulder abduction, exorotation and hand-to-mouth movement just before operation and the maximum result in 54 children

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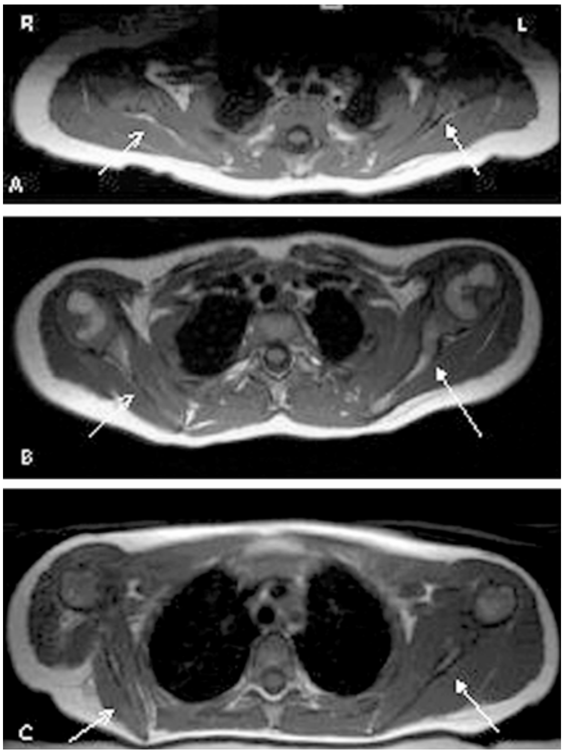

Figure 5

MR of the shoulder at the level of the infraspinatus muscles (arrows) in a child lacking spontaneous recovery of exorotation.

A) At 4 months of age,
B) At 3.5 years of age, one day before accessory to suprascapulare nerve transfer and
C) at 5.5 years of age, two years after surgery

In **A** the size of the infraspinatus muscle on the affected, right side (open arrow) is comparable to the left side.
In **B** there is some muscle wasting but not as pronounced as two years after surgery (**C**), when active exorotation has recovered.

MRI of the shoulder muscles (Fig 5) In three patients MRI of shoulder muscles showed less but still considerable muscle volumes of spinatus muscles on the affected side with some atrophy as compared to the non-affected side in pre-operative investigations. At two years follow-up after accessory nerve to suprascapular nerve transfer, muscle volume diminished with more extensive muscle wasting as compared to the non-affected side.

DISCUSSION

In children with an OBPL, who mainly fail to recover active shoulder exorotation, indication, timing and method of operative treatment to restore this function is a matter of debate (9, 17, 18, 28). Tendon, muscle or nerve transfers are treatment options (17, 25, 26, 28). In this study of 54 children we choose to perform an XIN to SSN transfer preferably between the age of 10 to 16 months. Evaluation of active exorotation as solitary movement is best performed conform to the description by Pondaag et al. (17). Commonly used scoring systems include other shoulder movements as well and do not describe exorotation specifically (4, 7, 8, 22). Functional results for exorotation and improvement of abduction were remarkable.
Simultaneous subscapular tendon lengthening to treat internal rotation contracture and to relieve passive deficit for exorotation in 14 patients was a prerequisite to allow regaining active exorotation to reach optimal individual results (25). As no active exorotation was present before operation, it is not a confounder in the evaluation.
As the nerve transfer does not interfere with other shoulder functions, we prefer to first perform nerve surgery to achieve an active motor function, targeting to the muscles most appropriate and meant by nature to effect this motion. Even older children have sufficient spinatus muscle volume and electro-myographic responses to justify this as a first procedure.
Alternate treatment as tendon and muscle transfers are only considered as a second option in children over the age of six years. Disadvantages are that original functions of muscles are sacrificed, that results of these procedures vary, and that long term results are disappointing (3, 9, 26, 28).

Functional recovery
Active shoulder exorotation in babies can be observed early in the first month and provoked in the third to fourth month as soon as they start to grasp and reach for subjects. Lack of active exorotation implies serious functional limitations for the affected upper limb. Activities such as eating, writing, dressing or combing hair may be hampered or impossible. To be able to put the hand to the mouth with adducted elbow, exorotation with a minimal range of -70 to 0 degrees is required. To compensate for the lack of exorotation the flexed upper arm will be abducted 90 degrees or more to reach the mouth (the so-called ‘Trumpet sign’, figure 1).
To be able to reach the back of the head some exorotation is required since this cannot be compensated by increased abduction and shoulder retraction. With prolonged lack of exorotation and in the presence of muscular imbalance, secondary endorotation

contractures and shoulder deformities, such as retroversion of the humeral head, may develop (23, 24). The objective to gain functional shoulder exorotation was obtained in 50 of 54 patients. The improvement of abduction that we observed in 50% of the patients can be explained by the contribution of the reinnervated supraspinatus muscle either or not in combination with a simultaneous, progressive increase in strength of the deltoid muscle. It could not be shown to what extent deficits of passive exorotation due to endorotation contractures and shoulder deformities relate to deficits in active motion or how passive deficits may restrict the range of active motion for exorotation.

The observed deficit for passive elbow extension in 15 patients is presumably due to a combination of a prolonged characteristic position of the arm in endorotation and slight flexion as well as progressive strength and dominance of elbow flexion over extension. With adducted shoulder, theoretically the long head of the triceps muscle can act as a weak exorotator but only when antagonized by forceful elbow flexion to eliminate its active elbow extension. Such a co-contraction is not observed and seems unlikely to explain the elbow flexion contractures.

Contradictory observations

Children showed spontaneous recovery of most functions of C-5 and C-6 or superior trunk within four months. The spinatus muscles showed some, but not extensive, muscle atrophy at preoperative physical examination and on MRI. There were no demonstrable signs of denervation on electromyography performed at four months of age and in the majority of patients intra-operative electro-stimulation of the SSN clearly elicited motor reactions in the infra- and supraspinatus muscles. In addition intra-operative electro-myography convincingly demonstrated normal muscle action potentials in five examined patients (fig 2), even in a child who previously underwent plexus exploration with transsection and grafting of the suprascapular nerve in the procedure of primary plexus reconstruction. These observations exclude a theoretically present second lesion in the SSN. Histologic examination of five investigated suprascapular nerves proved normal (fig 2). Because all these observations argue in favour of axonal continuity and integrity it is poorly understood why children fail to recover any spontaneous voluntary active shoulder exorotation.

Our results seem contradictory to the reported disappointingly low range (28% over 0 degrees) of active exorotation achieved when the XIN to SSN transfer is performed as part of a primary reconstruction of C5 and C6 lesions at an earlier age of around 5 months (17). Although we did not perform statistics on a similar group of infants, we also observed the tendency not to develop satisfactory results for active exorotation in these children. This observation actually was the reason to consider not to include the XIN to SSN transfer in the primary plexus reconstruction, but to restrict primary reconstruction of the SSN to a graft from C5 or C6 and reserve the nerve transfer for a separate secondary procedure if exorotation failed to recover within a year follow up. In the series of Pondaag et al. the severity of injury in children with a C5 and C6 lesion, meeting criteria for primary plexus reconstruction can be considered more serious than in the majority of the children in this study (17). Nevertheless, in those six children who also underwent primary surgery, there is a similar effect of the severity of the primary lesion and in addition a second denervation consequent to the nerve transfer. Although reliable statistics cannot be performed due to the small number and short follow up there may be a tendency not to reach values over 40 degrees. However, values over 0 degrees, already reached in four of six children, are functional and considered a major improvement for a child. In the series reported by Pondaag et al. the nerve transfers were performed up to 7 months of age. Effect of age was not studied extensively. Either young age of the nerve transfer, and /or the combination with more severe lesions and /or more extensive plexus reconstruction are factors that could negatively affect outcome of shoulder exorotation.

Defective motor programming in early infancy

From the moment infants first reach out to touch and grab, reaching patterns at first are tortuous and indirect with velocity peaks and bumps, periodic plateaus and even regression in control until a reliable reach trajectory control is developed around the seventh month (27). Each infant must discover its own solutions in relation to the given muscles, energetic levels and tasks at hand in conjunction with hand-eye coordination. Infants are both exploiting and modulating force characteristics of the limbs. Thus a kind of motor memory, encoding spatial form and speed changes, for a specific trajectory, becomes integrated. A central plan cannot be separated from the execution as the periphery sets the stage for central nervous changes. In infants with an OBPL and deficient peripheral tools to execute tasks within the trajectory of exorotation, the individual deviant solutions are cortically integrated during a presumably critical period until the seventh month of age. Despite intensive conservative efforts to stimulate a normal exorotation movement pattern the aberrant central plan does not seem to adapt to changes after this period. Most patients followed in our study group were well over one year of age and even contained 14 patients over two years of age, but still lacked any signs of active exorotation. The remarkable brain plasticity of infants in itself may be responsible for the setting of the disorderly motor program. Within the trajectory for exorotation, visually guided reaching patterns develop but are dysfunctionally set and executed. In stead of using the infra-spinatus muscle, which in potency is capable of function, reaching is altered and, as it seems irreversibly, provided by shoulder abduction and elevation in combination with elbow flexion and extension. Persisting lack of active exorotation can be considered as a form of developmental apraxia caused by defective motor programming with impaired motor unit activation of otherwise intact spinatus muscles (5). If the XIN to SSN transfer is performed as part of a plexus reconstruction, a comparable situation evolves where all shoulder functions have to be regenerated and centrally reintegrated simultaneously. For the same reason (developmental apraxia) this may have a negative effect on active exorotation. Young age may be a negative factor. The majority of the children studied, showed spontaneous recovery and early operation was therefore not justified. The effect of the XIN to SSN transfer when performed early (under 7 months), as single procedure is not studied. However, it seems well advised to wait, over 10 months, for regain of control of other shoulder functions, before trying to restore exorotation.

Type of lesion

Theoretically the primary type of nerve lesion in the studied patients, except in those who underwent early primary plexus reconstruction, could not have lead to extensive axonal loss and Wallerian degeneration (4, 11, 21) as most superior trunk functions recovered early, and patients did not show pronounced secondary muscle wasting at physical examination and on MRI studies. Absence of denervation of spinatus muscles on electromyography at four months in 39 of 40 patients might be another argument, although denervation in infants is known to disappear after 60 days anyhow (6). The observed integrity and reactivity of suprascapular nerves and spinatus muscles is likely to be preserved by spinal reflex activity to proprioceptic afferent signals or, involuntarily co-contractions with other muscles as a result of a disorderly central motor program (5, 6, 19). Axonal misdirection of outgrowing, regenerating axons is unlikely considering the presumed lesion types in our study group. Only in partial or complete neurotmesis at cervical root or truncal level misrouting into the suprascapular nerve with consequently ineffective reinnervation of target organs resulting in co-contraction with other muscles or ineffective outgrowth of mainly sensory axons into the target muscle may occur. Reactivity of spinatus muscles after intra-operative electro-stimulation of the supra-scapular nerve would also be present in such cases, but without actual voluntary muscle action. However, there would be more pronounced muscle wasting around the fourth month. A dramatic loss of motor neurons in the spinal cord is a fundamental feature after a peripheral nerve injury in newborns (1). With a diminished motor neuron population structural and functional integration of emerging connections between descending corticospinal tracts, interneurons, muscle afferents and motorneurons is impaired. However, motorneurons of uninjured sites may take over function of injured sites (10). In C-5 and C-6 lesions (Group I), especially the C-7 motorneuron pool can fulfil this task. Changes in spinal cord architecture in our Group I patients, may have been in favour of early recovery of other C5 and C6 functions rather than exorotation. Possibly dominantly exploiting the intact extensor functions of C-7 at first, followed by progressive gain of elbow flexion and shoulder abduction at the expense of suprascapular nerve function in the exorotation trajectory.

Plasticity central in recovery after the nerve transfer

Failure of conservative treatment justified the operative procedure as described, although many would hesitate to sever a seemingly intact, reactive nerve. As a direct result of sharp severing of the suprascapular nerve in the procedure of the nerve transfer, muscle wasting of spinatus muscles definitely occurs. Then spinatus muscles are denervated and begin to atrophy (11,21). Wallerian degeneration followed by axonal regeneration eventually lead to reinnervation of these muscles (11, 21). Even, although active exorotation has long since recovered, MR-scans at two years follow-up still demonstrate muscle wasting. For understanding the remarkable recovery of exorotation following the accessory to suprascapular nerve transfer, it can be hypothesized that by disconnecting the suprascapular nerve from its central disorderly plan and connecting it to another nerve and thus its representative cortical area, shoulder exorotation is granted cortical reorganization using again the extraordinary plasticity of the young brain, following an analogue process as described in adults for intercostal to musculo-cutaneus nerve transfer (2, 12, 14). Inhibitory influences of the original aberrant motor plan may thus be abolished. Investigation of cortical representation or its changes after peripheral nerve injuries and nerve transfers with techniques such as functional MRI, Magneto-Encephalography or transcranial magnetic stimulation is intriguing but probably not feasible in non-cooperative infants. If the development of normal exorotation patterns, is hindered by early ineffective cocontractions with, or dominance of, other muscles, the identification of these disorderly functioning muscles with neurophysiological studies may be useful. Injection of botulinum toxin and temporary elimination of selected muscles might facilitate active exorotation to develop in the motor program (19). Nerve transfers, as described, may thus be precluded and prevented.

CONCLUSION

In infants with a Group I OBPL and satisfying spontaneous improvement of all functions except shoulder exorotation a XIN to SSN transfer, performed as a separate procedure at ages over 10 months proved effective and safe to regain functional exorotation in the large majority of patients in this small study group of 54 children. The failure to develop voluntary active exorotation is thought to be the consequence of secondary, disorderly development of central nervous system connections and motor programs, rather than lack of potentially functional peripheral tools as the suprascapular nerve and spinatus muscles.

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CHAPTER 7

ENDOSCOPY ASSISTED SURAL NERVE HARVEST IN INFANTS

Based on:

Child’s Nervous System 1999;15:192-195

Endoscopy-assisted sural nerve harvest in infants

ABSTRACT

A minimally invasive method of endoscopy-assisted sural nerve (SN) harvest in infants with obstetric brachial plexus lesions requiring nerve grafting procedures was applied to reduce the skin incision size and scarring at the donor site. Endoscopic visualization was achieved using a flexible and steerable Neuroview neuronavigational endoscope (Promedics, NL) 2.3 mm in diameter and 12 or 18 cm long in a peelaway sheath (700-9F) attached to a video camera. Through three 1.5-cm skin incisions the SN could be dissected free using a 2.5-mm diameter nerve stripper, pituitary curette or pituitary scissors under endoscopic vision from the opposite direction. To prevent any central nociceptive pain behaviour the sural nerve was blocked by lidocaine, and sectioned first proximally in the popliteal fossa then distally at the lateral malleolus.

INTRODUCTION

Infants with severe obstetric brachial plexus lesions who do not achieve a spontaneous recovery are candidates for nerve grafting procedures, which are generally performed when they are 4–6 months of age [2, 5, 12]. The sural nerve (SN) is the best suited to use as an autogenous graft, because it is long, runs a straight course with minimal branching, and is readily accessible and entirely sensory, supplying a relatively unimportant dermatome [11]. In infants the SN is fragile and vulnerable. Harvesting techniques with blind use of nerve or tendon strippers or with multiple incisions in the calf are suitable for adults but may injure the nerve in infants [7, 8, 10, 13]. SN harvest in infants is usually performed by a “stocking-seam” incision over the calf [1], a safe and quick procedure. The nerve can easily be explored over its entire length, and if preferred the communicating branch of the peroneal nerve may be harvested in addition. However, the resulting scar is cosmetically difficult to accept (Fig. 1a). It has a tendency to thicken and widen and sometimes to develop keloid changes. Furthermore, these scars can cause an equinus position of the foot through adhesion with the Achilles tendon, as observed once in the author’s (unpublished) series of 60 children after open SN harvest.

As endoscopy has become part of the armamentarium of pediatric neurosurgeons [4, 6] this technique has been applied to assist in harvesting SNs in infants. When an endoscope is used it is possible to harvest the SN through three small (1–1.5 cm) incisions, with minimal scarring at the donor sites (Fig. 1b).

Fig. 1
a Hypertrophic donor-site scar after “stocking-seam” incision **b** Three 1.5-cm scars after endoscopic sural nerve (SN) harvest

ANATOMY

The SN is formed by the union of the medial sural cutaneous nerve and most often a single communicating ramus of the peroneal nerve. The medial cutaneous nerve (often synonymous with the SN) arises from the tibial nerve in the popliteal fossa and runs straight down between the bellies of the gastrocnemic muscle under the fascia, which it pierces in the middle to distal third of the calf, where it unites with the communicating ramus of the peroneal nerve. This branch runs down medially from the peroneal nerve under the fascia and over the lateral head of the gastrocnemic muscle [1, 8, 10, 11].

TECHNIQUE

The patient is positioned supine. Using a sterile diaper, both legs are prepared from the groin downward. After exploration of the brachial plexus the size and number of grafts needed to perform a reconstruction are estimated.

With the leg held straight at the knee and flexed and in abduction at the hip, a longitudinal incision 1–1.5 cm in length is made posterior to the lateral malleolus. The SN is identified, separated from the lesser saphenous vein and slung with a vessel loop. The next longitudinal incision is made in the midcalf. The SN is identified and labelled with a vessel loop. For identification it may be helpful to move the SN gently at the ankle. The third incision is made in the popliteal fossa, where the sural nerve is identified subfascially between the bellies of the gastrocnemic muscle. In all three incisions lidocaine 1% blocks are accomplished around the SN. Under direct vision, using operating loupes (Zeiss G3, 3 magnification) the SN is dissected free as far as possible (usually 2 cm) by cutting the overlying fascia away from the upper two incisions using Jameson's or pituitary scissors (Fig. 2).

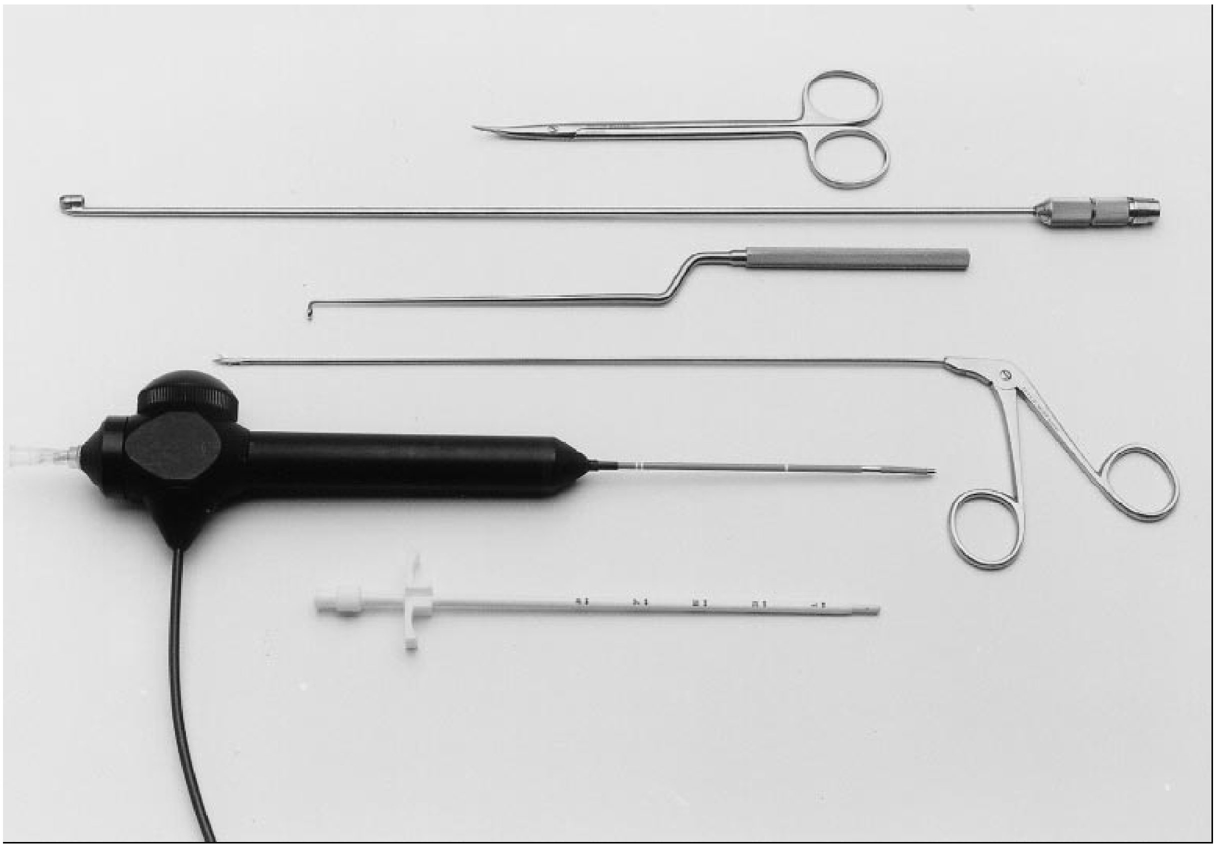

Fig. 2 Instruments used in endoscopic SN harvest. From *top* to *bottom*: Jameson's scissor, Asmuss nerve stripper (Aesculap, Germany), Hardy pituitary curette, straight 16.5-cm Nicola pituitary scissor, Neuroview neuronavigational flexible endoscope, 2.3 mm diameter, 12 cm length, and peelaway sheath, 700-9F (Promedics, The Netherlands)

To prevent multiple central pain reactions it is preferable to section the SN first close to its origin from the tibial nerve in the popliteal fossa and then distally at the lateral malleolus. After the preparation of small subcutaneous or subfascial pockets in both directions from the midcalf incision, a disposable flexible and steerable Neuroview neuronavigational endoscope (Promedics, The Netherlands) 2.3 mm in diameter and 12 or 18 cm long is introduced in a corresponding peelaway sheath (700-9F). The scope is advanced to just 0.5 cm or 1 cm from the tip of the sheath to prevent blood or fatty tissue from obscuring the operator's vision. It is quite easy to advance the scope in the sheath from the midcalf to the ankle incision over the SN with good visualization of the SN (Fig. 3). A 2.5-mm-diameter nerve stripper or a pituitary curette is advanced around the cut distal SN in a proximal direction, with simultaneous retraction of the scope in the subcutaneous or subfascial tunnel (Fig. 4a).
When tethering of the nerve is too tight to allow dissection in this way, the SN can be freed by introducing a straight pituitary scissor at the opposite side from or parallel to the scope. The distal part of the SN is then brought through the midcalf incision by gentle traction. The scope in the sheath is introduced in proximal direction from the midcalf following the SN under vision between the bellies of the gastrocnemic muscle to the popliteal fossa. Using the same procedure as already described, the SN is dissected free from the popliteal fossa in distal direction during retraction of the scope (Fig. 4b). Finally, the SN graft is delivered through the midcalf incision. There is no need for skin retraction during the endoscopy. Either the flexible tip of the endoscope or the opposed dissecting instrument can be used to create sufficient visual space for safe manipulation of the SN.
If the operator prefers to harvest the communicating branch of the peroneal nerve in addition, then the fascia should be incised from the midcalf incision in a proximal and lateral direction and a small brain spatula (5 mm wide) should be used for retraction, to create space for the endoscope over the lateral head of the gastrocnemic muscle. After a proximal lidocaine block, the nerve can be severed proximally with a pituitary scissor parallel to the endoscope. The skin is closed in two layers with absorbable intracuticular sutures. Postoperatively pressure stockings are used until the scars turn pale.
The size of an SN graft harvested either by open or by endoscopy-assisted dissection ranges from 10 cm to 13 cm in infants aged 4–6 months.

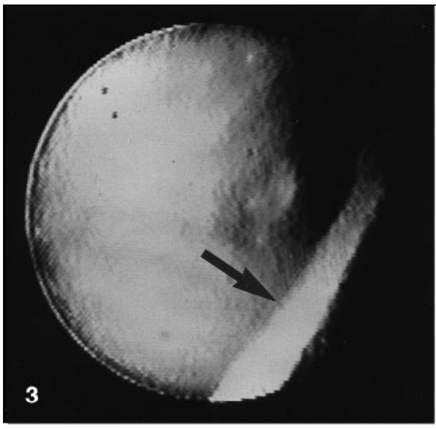

Fig. 3 Endoscopic view of SN (*arrow*) in subcutaneous tract in the distal calf

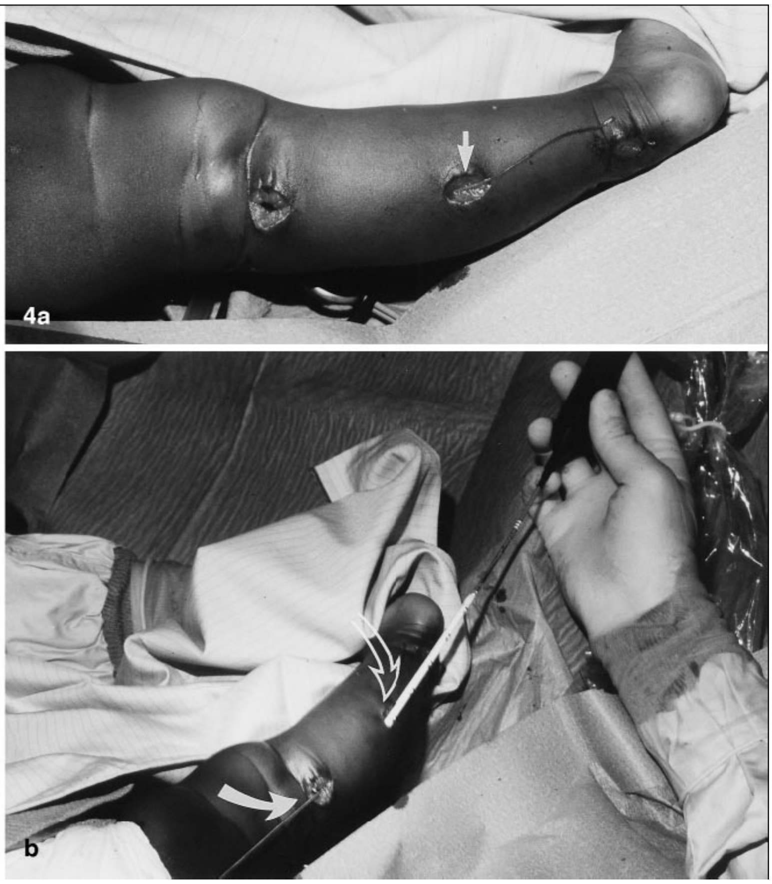

Fig. 4 **a** SN is first freed distally and delivered through the midcalf incision (*arrow*), and then **b** the endoscopic procedure is applied in the proximal calf, using the endoscope in a peelaway sheath from one side (*open arrow*) and a nerve stripper in the opposite direction (*arrow*)

In five patients eight sural nerves were harvested. Inspection of the nerve grafts under the operating microscope showed no injuries. Endoscopyassisted procedures take 35 min to 1 h.

DISCUSSION

Sural nerve harvest in infants requires long incisions when open techniques are used. The resulting scars are often hypertrophic and cosmetically not acceptable; they may cause functional disability with equinus position of the foot. Parents are shocked when informed of the size of the incisions necessary in the lower legs. Children are stigmatized for life by ugly scars, which may have negative influences on their psychosocial development.
Endoscopic instruments have broadened the range of therapeutic options open to neurosurgeons [4, 6]. In peripheral nerve surgery performed in pediatric patients endoscopy enables surgeons to perform minimally invasive procedures [1]. The main advantage of endoscopy-assisted SN harvest is that visible scarring is remarkably reduced. Earlier reports have described SN harvest under endoscopic visualization [1, 9]. Our technique is different in that no tourniquet control is used. No bleeding that interfered with endoscopic vision was experienced. Introduction of retractors [1] or inflatable balloon catheters [9] to create relatively large subcutaneous or subfascial spaces could be avoided by using a peelaway sheath and a flexible, steerable endoscope introduced from one direction and the other instruments described from the opposite direction.
If endoscopic dissection should fail and an open procedure be required, three longitudinal incisions that can be connected later are preferable to transverse incisions along natural skin lines.
The operating time of 1 h at the most was acceptable compared with the about 25–35 min needed for open procedures.
The importance of lidocaine blocks, especially around the proximal nerve before it is sectioned, and of first cutting any nerve proximally is stressed. There is clinical and experimental evidence showing that any neurectomy produces central changes that influence nociceptive pain behaviour after nerve sections [3, 14]. In amputees phantom pain and in rats autotomy behaviour were dramatically reduced when nerves were blocked with local anesthetics before they were sectioned. Although there are no reports on pain behaviour related to SN harvest in children, it is recommended that the precautions mentioned be observed.

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CHAPTER 8

DISCUSSION

DISCUSSION

Obstetric brachial plexus lesions: Aspects of Diagnosis

The incidence of OBPL in general is not decreasing over the past decennia. Aspects of diagnosis and treatment will continue to be in focus for further research. The literature is overly optimistic in stating that over 80% of cases with OBPL will show ‘good’ spontaneous recovery. A recent critical outcome study showed only 66% complete recovery (Hoeksma, 2004). This underlines the potential for lasting functional deficits in OBPL and the importance of carefull, long term follow up in dedicated centres.

In a newborn with OBPL of no matter what severity it is not possible to predict the course of recovery. The question to raise is: “what to do when and when to do what ?”

In several studies published in the literature this question was the central issue and some anwers are formulated.

In severe OPBL, the best indicator for further treatment decisions is biceps function at three months (Tassin 1984, Hoeksma 2004). When children are able to flex the elbow and to put the hand into their mouth against gravity, further conservative treatment is justified. However, by no means one is allowed to conclude that recovery will be complete. If function of the biceps has sufficiently recovered at three months, then further prediction of neurological outcome is best based on active shoulder exorotation and forearm supination (Hoeksma e.a., 2004).

If, on the other hand, there is no sign of recovery of the biceps muscle at three months the overall outlook for recovery of shoulder and arm function is gloomy and neurosurgical exploration is indicated. The indication for neurosurgical plexus exploration and reconstruction is thus only based on clinical follow up studies of neurological and functional recovery. Needless to say, that referral of infants with an OBPL should be before the age of three months.

If the decision to perform surgery is made at three months, preoperative ancillary investigations should ideally delineate the extent and type of the nerve lesion, as described in chapters 2, 3 and 4. Reconstruction with nerve grafts is based on intrinsic, regenerative capacity of nerve stumps. Avulsed roots have no regenerative capacity. When during operation a seemingly intact nerve root at a neurofaramen is used as a nerve stump in reconstruction with nerve grafts, but is actually avulsed from the spinal cord, axonal regrowth is impossible and recovery of this nerve function remains an illusion.

As described in chapter 3 of this thesis, there are different techniques for electrodiagnosis and diagnostic imaging. Results of different electrodiagnostic tests are especialy difficult to interpret in babies for a number of reasons. Regarding the actual type of a lesion, the combination of electromyography and nerve action potential studies will potentially give the best possible information on the presence of a conduction block or avulsion.The first allows an expectative attitude, the other urges surgical exploration. In general, preoperative electrodiagnosis offers too optimistic results and is almost always in conflict with a concerning clinical condition. New magnetic resonance imaging techniques enable visualization of plexial structures from the spinal root entry zones to the level of the peripheral plexial structures. For imaging the intraspinal trajectory of nerve roots different techniques are more appropiate than for the peripheral nervous parts. In spite of the newly described 3D-CISS MRI technique (chapter 3 and 4), especially suited for imaging the intraspinal trajectory of cervical nerve roots, some uncertainty remains as to the correct diagnosis of complete or incomplete avulsion of anterior and posterior roots. Unfortunately there is no gold standard to determine root continuity other than by direct surgical inspection necessitating laminotomy which is not applicable in infants.

Aspects of treatment

The failure to develop active exorotation.

A number of children, that were not elected for surgery at the age of three months will nevertheless end up with incomplete neurological recovery. A problem, addressed in chapter 5 of this thesis, is developmental apraxia as apparent cause of lack of improvement of active shoulder exorotation in a substantial number of otherwise spontaneously recovering children. Allthough individual muscles may have normal volumes on clinical and MRI examination and show voluntary movement at electromyographic studies, moreover direct stimulation of the suprascapular nerve effects muscle contraction, actual muscle performance may be absent. In fact the peripheral tools, muscle and nerve, appear ready to use, yet central changes prohibit development of movement. Paralysis of shoulder and arm muscles at a critical time of development of motor programmes may apparently result in apraxia.

Performing a nerve transfer from the accessory to the suprascapular nerve as a separate procedure at ages over 10 months, active functional shoulder exorotation is achieved in 52 of 54 patients. To exclude contribution of glenohumeral movement in the assessment of active exorotation, the exorotation should be scored with a flexed elbow and the upper arm in mildly fixed adduction. Most publications use scoring systems in which contribution of scapula retractors obscures pure exorotion by infraspinatus muscle activity. The majority of children in this study actually showed active function of these possibly obscuring C5 innervated muscles as the deltoid for shoulder abduction and rhomboids for scapular retraction. However, the active exorotation achieved, as described in *chapter 5,* is the result of infraspinatus muscle activation after reinnervation of the SSN by transfer from the accessory nerve. This nerve transfer proved to be the missing link in the chain of central to peripheral nervous components necessary to execute shoulder exorotation.

Concluding in this study, that development of central nervous system motor programming is not adapted to execute shoulder exorotation, it will be a future challenge to develop methods to study the relevant central nervous system changes before and after succesfull treatment. Dynamic imaging techniques are promising in showing central nervous system activity in relation to movement. These techniques require cooperation of patients and can only be applied in selected older children. A protocol to apply magnetencephalography is currently being prepared at VU University Medical Center.

Secondary deformities of the shoulder

Children with an OBPL are at risk to develop characteristic secondary shoulder deformities consisting of a flexion and internal rotation contracture of the shoulder as well as a posterior subluxation of the humeral head. MRI studies have shown early changes becoming more pronounced with age in the still mainly cartilaginous shoulders in infants.

The deformities are possibly a consequence of muscular imbalance of shoulder muscles, with dominance of adductors and internal rotators. Functional implications are unclear in early phases, whereas pronounced late deformities with humeral head subluxation and severe glenoid deformities at least will affect passive range of movement.

Depending on the recovery of neurologic deficits, as yet, elective orthopedic surgical treatment of secondary shoulder deformities is part of the integral multidisciplinary treatment program.

Prevention of secondary shoulder deformities is an issue to discuss. Counteracting muscular imbalance of shoulder muscles by applying botoxuline to adductors and internal rotators in the early phase of recovery, for instance between the fourth and eight month, may prevent these deformities but may cause other, unforeseen deformities as well. In the

Group I patients described in *chapter 5*, botoxulinum toxin may simultaneously allow spontaneous development of active exorotation. But there is a risk of negative effects on the development of motor programs for internal rotation.

Endoscopy assisted harvest of the sural nerve.

The sural nerve is the most widely used donor nerve in grafting procedures. As not to harm the delicate nerves, large longitudinal or sinusoid skin incisions were in vogue to explore and harvest the sural nerves in their entire length. As a consequence, resulting scars were ugly and often caused functional limitations due to skin contractures. With the technique described in *chapter 7*, sural nerves can be harvested through three small, cosmetically acceptable and functionally non complicating skin incisions. Wheras advantages of the technique are obvious, moreover the surgeon experiences in these procedures, that sural nerves can even be harvested without the use of an endoscope. The three small incisions allow direct subcutaneous and subfacial vision of three centimetres in both directions, which is sufficient to control and preserve the integrity of the nerve during preparation along its trajectory from the kneefold to the ankle. Even if anatomical variations are encountered, such as contributions from both tibial and peroneal nerves. Meanwhile, in the kneefold skin incisions are modified and placed in the transverse, natural direction of the fold to prevent skin contractures even after small but longitudinal incisions. The technique as descibed in the thesis, thus formed the basis of another modification to harvest the sural nerve in infants not yet described.

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SUMMARY

A summary is made up of this thesis on aspects of diagnosis and treatment in obstetric brachial plexus lesions (OBPL).

In **CHAPTER 1** the general outline of this thesis is described. The aim of this thesis was to study aspects of diagnosis and treatment in OBPL. For this purpose several specific objectives were formulated. First, to make an inventory of and select, develop or modify ancillary investigative tools that will contribute to the most likely correct diagnosis of OBPL and possible secondary deformities,

Secondly, to evaluate the effect of a relatively easy to perform nerve transfer in carefully selected patients with a typical clinical picture lacking active shoulder exorotation.

Thirdly, to improve the surgical technique to harvest the sural nerve, the most commonly used donor nerve, by developing a minimally invasive procedure.

In **CHAPTER 2** an overview of the etio-pathology, epidemiology, diagnosis, prognosis and treatment of OBPL is given. The Narakas classification, a clinical classification method of the severity of the OBPL is presented. The prognosis of OBPL is usually good with the majority of infants showing a near to or full recovery within the first year. In a minority of infants, neurosurgical reconstruction of damaged plexial structures is indicated because of the severity of the nervous lesion. Since root avulsions from the spinal cord demand a special operative strategy the purpose of additional investigations is to diagnose this type of lesion. Outcome measurement methods (e.g. the Mallet score) for the evaluation of treatment results are presented.

In **CHAPTER 3** an overview is presented on current electrophysiological and imaging techniques as ancillary, preoperative, investigative tools to determine the type, level and extent of the nerve lesion in OBPL. Results of electromyography (EMG) should be differently appreciated as compared to adults. More often than not, in infants with OBPL, there is a discrepancy between disapointing lack of actual functional muscle activity and optimistic EMG findings with motor unit potentials (MUP’s). Study of nerve action potentials and, more so, combined with EMG, differentiation may be possible of a nerve conduction block or root avulsion. Somatosensory evoked potentials, at the best, only offer information on electrical conduction in sensory roots. Motor evoked potentials with direct intra-operative registration at cervical roots would theoretically be very informative on conduction in motor roots, but is not applicable in infants (yet).

For imaging brachial plexus structures in detail in infants magnetic resonance imaging (MRI), applying different techniques in a non-invasive way, has surpassed other imaging techniques. Especially roots can accurately be traced to the spinal cord, thus allowing diagnos of avulsions with increasing reliability.

In **CHAPTER 4** a new technique is described for imaging the intraspinal trajectory of spinal nerve roots. In this study it is applied to demonstrate root avulsions of cervical root C7 in ten infants with an OBPL mainly affecting the muscles innervated by C7. With this 3D-constructive interference in steady state MRI anterior as well as posterior roots can be traced in their intraspinal course. Intra-operative findings matched the predicted lesions based on preoperative MRI studies in the majority of patients. However, exact prediction on the presence of a root avulsion still was not possible in a few cases. The value of routine EMG in this study was restricted as a supportive tool in operative decision making.

In **CHAPTER 5** findings are presented of a prospective study on early secondary deformations of the shoulder in infants with severe OBPL, who showed insufficient spontaneous recovery at three months of age. Using a standarized MRI-protocol for the shoulder in 26 patients (mean age of 5.6 months). Measurements were made of the glenoid, glenoid version and position of the humeral head. The appearance of the glenoid on the affected side was normal in only 11 shoulders. In the remainder it was convex in eight and biconcave in seven cases.The degree of humeral head subluxation was significantly greater ($p=0.001$) in affected shoulders than in normal appearing shoulders (152 and 170 degrees, respectively). The presence of abnormal glenoid retroversion and humeral head subluxation increased with age: there was a statistical difference ($p=0.001$) between infants younger than 5 months and those who were older. As yet functional implications of early shoulder deformities are not clear.

In **CHAPTER 6** a study is described on the effect of an accessory to suprascapular nerve transfer, performed as a separate procedure, to restore active shoulder exorotation. In 54 children with an OBPL who otherwise showed satisfactory recovery but only lacked active shoulder exorotation, active movement was restored and improved from –70 degrees to functional levels over 0 degrees exept in two patients. Shoulder abduction also improved in the majority of infants. Because most other plexial functions in these infants spontaneously recovered, intra-operative electrostimulation of suprascapular nerves elicited spinatus muscle reaction, histology of suprascapular nerves proved normal and spinatus muscles did not show wasting on preoperative examination or MRI, a peripheral nerve lesion as main cause for the deficit is unlikely. Central nervous changes with consequent developmental apraxia may be principal causes for the lack of spontaneous recovery of active exorotation.

In **CHAPTER 7** a minimally invasive operative technique to harvest the most commonly used donor nerve in grafting procedures, the sural nerve (SN), is described. Using a flexible 2.3 mm in diameter endoscope attached to a video camera, it was possible to dissect the SN in the subcutaneous and subfacial tissue of the calf. Through three 1.5 cm skin incisions and using a nerve stripper, pituitary curette or pituitary scissors opposed to the endoscope SN's can be harvested. Large skin incisions and consequent ugly, often contracting scars can thus be avoided.

CHAPTER 8 Various aspects of diagnosis and treatment of OBPL as described in the previous chapters are discussed and conclusions are formulated. Because the incidence of OBPL does not decline there will be a continuous demand for multidisciplinary treatment and research in specialized centres. Decisions for conservative or operative treatment are made around the fourth month and are based on evident, spontaneous recovery of biceps function. Electrodiagnostics have limited value in the preoperative work-up of patients, whereas new MRI-techniques enable ever improving high resolution imaging of cervical nerve roots and possible types of traumatic lesions, as well as of early secondary deformities of the shoulder joint. Lack of spontaneous recovery of shoulder exorotation in otherwise recovered children is most likely due to developmental apraxia and thus may be caused by aberrant adaptation of the central nervous system in response to a brachial plexus lesion. If tested, the suprascapular nerves and spinatus muscles are potentially capable to function, however not voluntarily used. An accessory nerve to suprascapular nerve transfer will effect functional exorotation in the vast majority of selected children. Further research is required to study central nervous changes in reaction to peripheral nerve lesions and/or surgical reconstructions.

Only three small skin incisions in the calf are sufficient to harvest the most commonly used donor nerve, the sural nerve to reconstruct a damaged brachial plexus. Endoscopy to assist harvesting is rarely indicated.

SAMENVATTING

Aspecten van diagnose en behandeling bij obstetrische plexus brachialis letsels.

HOOFDSTUK 1 Dit hoofdstuk biedt een overzicht van de in dit proefschrift te beschrijven onderwerpen. Een aantal aspecten van diagnose en behandeling van obstetrische plexus brachialis letsels (OPBL) zijn beschreven of vormen onderwerp van verdere studie. Eerst is een inventarisatie gemaakt van verschillende methoden van klinisch onderzoek en zijn de meest gangbare diagnostische hulpmiddelen kritisch geëvalueerd. Vervolgens zijn een aantal studies verricht, die apart beschreven worden. Nieuwe beeldvormende methoden van onderzoek werden ontwikkeld en getest. Een in het VU medisch centrum ontwikkelde beeldvormende techniek, MRI-scan, maakt het mogelijk om zenuwen scherp af te beelden vanaf de oorsprong in het ruggenmerg tot waar de zenuw het halswervelkanaal zijdelings verlaat. Met een andere techniek kunnen veranderingen in vorm en stand van het schoudergewricht bij baby's afgebeeld en gemeten worden. Het bijzondere hiervan is, dat vooral veranderingen in de vorm van het kraakbeen, waaruit de schouder van een baby eigenlijk nog is opgebouwd, goed zichtbaar gemaakt worden.
Bij een aantal kinderen met een onvermogen de arm in de schouder naar buiten te draaien, ofwel te exoroteren, werd een zenuwoperatie verricht, waardoor de kinderen, na verloop van tijd, in staat waren deze beweging uiteindelijk toch te kunnen maken.
Bij operaties van de plexus brachialis is het meestal nodig verscheurde zenuwen te reconstrueren met behulp van zenuwtransplantaten. Hiertoe wordt veel gebruik gemaakt van de kuitzenuw, de Nervus Suralis. Een techniek wordt beschreven om deze zenuw via drie minimale huidsneden en met behulp van een endoscoop (kijkoperatie) uit de kuit te verwijderen.

HOOFDSTUK 2 Een obstetrisch plexus brachialis letsel is een geboortetrauma, waarbij zenuwen, in het verloop van het halsruggenmerg en het gebied in de schouder boven het sleutelbeen, verscheuren of zelfs uit het ruggenmerg losgetrokken worden. Electrische geleiding in de zenuw valt weg, waardoor spieren in de schouder en arm verlamd raken. In dit hoofdstuk wordt een overzicht gegeven van ontstaansmechanisme, epidemiologie, diagnose, prognose en behandeling van het OPBL. De indeling naar ernst en uitgebreidheid van OBPL volgens Narakas is weergegeven. De meeste OPBL zijn niet ernstig en de prognose voor nagenoeg volledig spontaan herstel is goed voor meer dan 80% van de kinderen. Een minderheid heeft een dusdanig ernstig zenuwletsel, dat operatieve reconstructie van de zenuwen van de plexus nodig is. Hiertoe wordt beslist, wanneer baby's op de leeftijd van drie tot vier maanden geen biceps functie hebben. Kinderen ondergaan dan een spiertest, electromyografie, waarmee wordt onderzocht of en hoedanig een spier door een zenuw geprikkeld en aangestuurd wordt. Tevens wordt een MRI-scan gemaakt om in beeld te brengen of een zenuw uit het ruggenmerg is los gescheurd. Tenslotte worden resultaten van operatieve behandeling en methoden van lichamelijk onderzoek ter beoordeling van schouder- en armfunctie beschreven.

HOOFDSTUK 3 Hier wordt nader ingegaan op de meest gangbare en toegepaste middelen van onderzoek om meer informatie te verschaffen over het type, de uitgebreidheid en de ernst van een OPBL . Zogenaamd neurofysiologisch onderzoek, gebruik makend van allerlei elektrodiagnostische methoden om functies van zenuwen en spieren te onderzoeken, geeft bij baby's volledig anders te interpreteren uitslagen in vergelijking met volwassenen. In het algemeen zijn de uitslagen van deze onderzoeken veel te optimistisch. Bij nagenoeg volledig functioneel verlamde spieren kan de uitslag van een onderzoek bij baby's maar weinig afwijkend zijn. Door verschillende technieken van onderzoek te combineren kunnen enkele typen letsels onderscheiden en vastgesteld worden. Een moeilijk op te lossen probleem is te weten te komen of een zenuw helemaal of maar gedeeltelijk uit het ruggenmerg is losgetrokken. Indien dit het geval is, kan een neurochirurg deze zenuw niet gebruiken om een zenuw te reconstrueren. Met zeer gedifferentieerd onderzoek is het wellicht in de toekomst bij baby's beter mogelijk om door een stroomstootje via de schedel de continuïteit van zenuwbanen van de hersenen tot in de spieren te meten (Motor Evoked Potentials ~ MEP) maar dan nog blijft de vraag of de kwaliteit van een zenuw goed genoeg is voor de vereiste functie.
Allerlei aanvullende beeldvormende technieken met MRI-scans geven steeds betrouwbaarder informatie over het type letsel van de verschillende aan de plexus bijdragende halszenuwen. Helaas kan een avulsie van een zenuw (uit het ruggenmerg losgetrokken zenuw) nog steeds niet met zekerheid worden aangetoond.

HOOFDSTUK 4 In het VU medisch centrum werd in samenwerking met de neuroradiologen een nieuwe MRI-techniek geëvalueerd. Met deze 3D-CISS-MRI kunnen de zenuwwortels, die aan de achter– en voorkant van het ruggenmerg in de hals geworteld zijn, afgebeeld worden. Bij een geselecteerde groep patiëntjes, waarbij vooral verlammingsverschijnselen passend bij letsel van de zevende halszenuw waren vastgesteld, bleek de bevinding van de MRI scan goed overeen te komen met de bevindingen bij operatie. Bij operatie herken je een avulsie, doordat het begin van een zenuw, dat twee takjes heeft, tot buiten de wervels weggetrokken is. Verraderlijk is het, wanneer het losgetrokken zenuwtje net niet buiten de wervels is terecht gekomen. Dan lijkt zo'n zenuw normaal en zou de operateur per vergissing op deze inactieve en kapotte zenuw een andere zenuw kunnen aansluiten. Een zenuwreconstructie is namelijk alleen mogelijk door gebruik te maken van zenuwen, die geworteld zijn in het ruggenmerg. Alleen dan behouden zenuwen het intrinsieke vermogen van regeneratie. De MRI biedt weliswaar extra informatie maar desondanks kan helaas nooit met 100% zekerheid een uitspraak gedaan worden over het al dan niet aanwezig zijn van een zenuw-avulsie. De toegevoegde waarde van routine neurofysiologisch onderzoek bij het oplossen van het gestelde probleem was gering.

HOOFDSTUK 5 Kinderen met een OPBL hebben een verhoogd risico op gewrichtsafwijkingen van de schouder. Sommige spieren rond de schouder zijn verlamd en andere juist zeer krachtig. Hierdoor is er een ongelijke krachtsverdeling rond het schoudergewricht. Tijdens de groei en ontwikkeling van het gewricht kunnen hierdoor vervormingen ontstaan. Bij 26 kinderen met een ernstig OPBL werd op een gemiddelde leeftijd van 5,6 maanden een MRI-scan van de schouder verricht. Bijzonder hierbij is, dat juist veranderingen in het kraakbeen, waaruit de schouder bij baby's nog is opgebouwd, kunnen worden aangetoond Er waren slechts 11 normale schouders. Bij 8 kinderen was de schouderkom te hol, terwijl er bij 7 een richel aangetoond werd. De kop van de schouder stond bij deze kinderen beduidend meer naar achteren verplaatst. Dit was des te meer uitsproken naarmate de kinderen ouder waren. De functionele betekenis van deze al vroeg aan te tonen afwijkingen van het schoudergewricht is nog niet duidelijk.

HOOFDSTUK 6 Op de leeftijd van vier tot zes maanden hadden 54 kinderen met een OPBL voldoende spontaan herstel van schouder en armfuncties. Een grote operatie met reconstructie van zenuwen van de plexus was daarom niet nodig. Met intensieve fysiotherapie verbeterden de schouder- en armfuncties weliswaar, maar herstel van één beweging bleef uit. Deze kinderen waren niet in staat de arm in de schouder naar buiten te draaien (exoroteren). Er werd minstens afgewacht tot de tiende maand, waarna alsnog een kleine zenuwoperatie verricht werd. Een takje van de zenuw voor de monnikkapspier (nervus accessorius) werd afgetakt en aangesloten op het begin van de zenuw (nervus suprascapularis), die de spieren aanstuurt, waarmee exorotatie verricht wordt (vooral de musculus infraspinatus). Bij 52 kinderen werd hierdoor een functionele actieve exorotatie bereikt. Het is moeilijk te verklaren waarom de meeste schouderfuncties wel spontaan herstellen, maar juist de exorotatie niet. Des te meer is dit verwonderlijk omdat de spieren voor de exorotatie niet atrofiëren, de zenuw (nervus suprascapularis) bij operatie goed elektrisch te stimuleren is, waardoor spierreacties geprovoceerd kunnen worden, en weefselonderzoek van de zenuw niet afwijkend is. De kinderen lijken hun intacte zenuw en spieren voor de exorotatie in de schouder gewoonweg niet te gebruiken. De mogelijke oorzaak is, dat kinderen met een OPBL tussen de derde en de zevende maand, een fase waarin bewegingen aangeleerd worden en er definitieve verbindingen tussen hersenen, ruggenmerg en perifere zenuwen gemaakt worden, abnormale bewegingspatronen aanleren. Het vermogen om daarna nog zelf het exoroteren te leren lijkt verloren te zijn gegaan. Door de toegepaste operatie te doen op een leeftijd wanneer de andere schouderfuncties redelijk zijn hersteld, wordt dit vermogen opnieuw verkregen, doordat een andere zenuw, die eigen verbindingen met de hersenen heeft, wordt benut.

HOOFDSTUK 7 Bij een ernstig OPBL is een operatie nodig om kapot gescheurde zenuwen te reconstrueren. In het verloop van een zenuw aan te treffen beschadigde delen en littekenknobbels worden verwijderd. Tussen de twee overblijvende zenuwstompjes kunnen stukjes donorzenuw aangebracht worden, waardoor de zenuw weer kan uitgroeien naar de verlamde spieren om deze uiteindelijk weer aan te sturen. De kuitzenuw (Nervus Suralis) is als donorzenuw zeer geschikt. Deze zenuw loopt onderhuids van de knieholte tot de buitenenkel en verzorgt het gevoel in een stukje huid over de hiel. Het was gebruikelijk om met een grote huidsnede van de knieholte tot de enkel, meestal in zigzagvorm aangebracht, de zenuw te verwijderen. Resulterende littekens waren cosmetisch akelig en gaven bovendien nogal eens nare contracturen. Door een nieuwe kijkoperatie, gebruik makend van een endoscoop, kan de zenuw via drie kleine huidsneden, in de knieholte, het midden van de kuit en bij de buitenenkel, evengoed verwijderd worden. De 2 à 3 cm littekens zijn fraai en functioneel niet beperkend.

HOOFDSTUK 8 Aan verschillende aspecten van diagnose en behandeling van het OPBL, zoals beschreven in voorgaande hoofdstukken, wordt een discussie gewijd. Omdat het aantal kinderen dat tijdens de geboorte een OPBL oploopt niet lijkt af te nemen, zullen gespecialiseerde centra zich blijvend moeten inzetten om de kwaliteit van behandeling te verbeteren. Onder meer door onderzoek te verrichten. De beslissing een kind met een OPBL al dan niet te opereren wordt rond de vierde maand genomen en is gebaseerd op het herstel van de spierfunctie van de biceps. Electrodiagnostiek heeft een beperkte waarde bij het preoperatieve onderzoek van kinderen met een opbl. Met moderne MRI-scans is het steeds beter mogelijk om zenuwen en zenuwletsels af te beelden. Helaas kan desondanks nog geen 100-procent zekerheid omtrent een avulsie van een zenuw geboden worden.
Bij spontaan goed herstelde kinderen met een OPBL, die alleen geen actieve schouder-exorotatie hebben, lijken de hiertoe benodigde zenuw (nervus suprascapularis) en spier (musculus infraspinatus) weliswaar intact, maar kunnen niet willekeurig gebruikt worden. Dit berust hoogst waarschijnlijk op een reactie van het centrale zenuwstelsel op het plexus letsel, met als gevolg een apraxie voor de exorotatie. Voor een beter begrip van deze reacties in de hersenen is verder onderzoek nodig. Door de nervus suprascapularis aan te

sluiten op een andere zenuw (tak van de zenuw voor de monnikkap spier) wordt het vermogen de schouder te exoroteren bij het merendeel van geselecteerde kinderen hersteld. Slechts drie kleine huidsneden in de kuit zijn voldoende om de kuitzenuw, die als donorzenuw bij zenuwreconstructies wordt benut, te kunnen verwijderen. Als hulpmiddel bleek een endoscoop nuttig, maar is eigenlijk zelden of nooit meer nodig.

PERSONAL REMARKS

During neurosurgical training in Germany I was introduced in the field of adult plexus surgery by Prof. Dr. M. Samii and his senior staff, in particular Prof. Dr. Goetz Penkert. Being appointed pediatric neurosurgeon at VU Medical Centre (VUmc) in 1992, the late neurosurgeon Dr. A.(Bart) C.J. Slooff, based in Heerlen, was a guide into and a continues stimulus to explore the field of OBPL. It was a privilege to have worked together and I keep precious memories of our joined ‘plexus tours’ to Linz in Austria. There we always enjoyed the warm hospitality of my Austrian colleague and friend Prof. Dr. Kurt Holl and operated on quite a number of children, superbly assisted by the multitalented nurse Michi Nimmervoll, who also provided data and slides for this thesis.

At VUmc integral treatment of children with OBPL seriously commenced in 1994. At present about 60 new cases each year are presented. Ever since, dynamic specialists are contributing to the multidisciplinary plexus team. From the start it was always a joy to do our weekly out patient plexus clinic together with enthusiastic colleagues from the department of rehabilation medicine. For many years (Prof. Dr.) Frans Nollet was my pal and I admire his accuracy, creative mind and sense of humor. He was a critical co-author of some papers contributing to this thesis. After he left VUmc last year, I enjoy the cooperation with the physiatrists Mirjam van Doorn, Vincent de Groot and Katinka Folmer. Our plastic and reconstructive surgeons, Marco Ritt and Hay Winters I owe gratitude for the always pleasant cooperation and their expert opinion on and superior craftmanship in the treatment of secondary functional problems. The pediatric orthopedic surgeon Hans van der Sluijs, who in his thesis broadened the insight in secondary deformities in OBPL, is a true companion. Not only in our Plexus Team at VUmc but also in the cooperation with other centres in Europe which we visit together. At present there is a bi-annual coöperation with the Landes Nervenklinik and the Kinderklinik in Linz, Austria and the Neurochirurgische Klinik of the Universitätsklinikum in Giessen, Germany, where operations are scheduled and at the same time friendship is shared with our colleagues Kurt Holl (Linz) and Andreas Jödicke (Giessen).

This thesis was only possible with the support of many colleagues, most of whom are co-authors of the papers contributing to this thesis. Special gratitude I owe to Prof. Dr. Frederik Barkhof who perfected the 3D-CISS-MRI technique (described in chapter 4), to Dr. Rob Strijers for always being present, accompanied by his charming ladies, to perform intra-operative neuromonitoring and for his critical contribution as co-author, to Dr Bernard Uitdehaag for his guidance through statistics. Treatment of OBPL is not possible without support of the departments of physiotherapy, occupational therapy and of our cast technicians and nurses from the ward and operating theatres.

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For all the work done and yet to be done to guide me through the defense of this thesis and its aftermath I thank my dear colleague and room mate Saskia Peerdeman and my friend Cees Klapwijk and wish them good luck.
Without the love and affection of my wife Doris and children Roelof, Mirjam and Nora life would not be so bright. As a pediatric neurosurgeon it is a great privilege to take care of children and many times when a baby, is presented at my out patient clinic I think of Louis Armstrong's song:

"I hear babies cry, and I watch 'em grow
They'll learn much more than I'll ever know
And I think to myself, what a wonderful world"

CURRICULUM VITAE

De auteur van dit proefschrift is geboren op 6 januari 1957 te Rotterdam. In 1977 werd het Atheneum B diploma behaald aan de Gereformeerde Scholengemeenschap te Rotterdam. Na aanvang van de studie geneeskunde in Antwerpen in 1977 werd deze een jaar later voortgezet aan de Erasmus Universiteit in Rotterdam waar in mei 1985 het arts examen met goed gevolg werd afgelegd. Door stages op diverse neurochirurgische afdelingen, in het Westeinde Ziekenhuis te Den Haag bij Dr. M.Th.A. van Duinen, het H. Verwoerd Hospitaal te Pretoria bij Prof Rensenbrink en het Universitätsspital te Zürich bij Prof. M.G. Yasargil werd tijdens de studie geneeskunde de interesse voor de neurochirurgie aangewakkerd. Na een eerste aanstelling als arts assistent chirurgie in het Bronovo Ziekenhuis te Den Haag werd van januari 1986 tot maart 1992 de opleiding tot neurochirurg gevolgd en afgesloten na het behalen van de “Facharztprüfung” bij Prof. Dr. Dr. hc. mult. M.Samii in het Nordstadt Krankenhaus te Hannover in Duitsland. Interesse in de kinderneurochirurgie werd tijdens de opleiding in Hannover gestimuleerd door één van de vaders van de kinderneurochirurgie (wijlen) Prof. Dr. Anthony Raimondi, die als visiting professor aan het Nordstadt Krankenhaus verbonden was. Sinds 1992 is de auteur als (kinder)neurochirurg aan het VU Medisch Centrum te Amsterdam verbonden. Eén van de zwaartepunten van de kinderneurochirurgie is de behandeling van kinderen met een obstetrisch plexus brachialis letsel, dat tevens het onderzoeksgebied werd.
De auteur is verder, onder meer, actief als President van de Scientific Committee van de European Society for Pediatric Neurosurgery en ‘editor’ van twee wetenschappelijke tijdschriften (Child’s Nervous System en Journal of Reconstructive Microsurgery).
De auteur is gehuwd met Doris Michel, en zij hebben drie fijne kinderen: Roelof, Mirjam en Nora.